Université de Montréal

Identification des facteurs sociocognitifs liés à l'adhésion au traitement des jeunes atteints d'hémophilie : une étude pour guider l'intervention en milieu hospitalier

par Sarah Bérubé

Faculté des Sciences et des Arts
Département de psychologie

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Identification des facteurs sociocognitifs liés à l'adhésion au traitement des jeunes atteints d'hémophilie : une étude pour guider l'intervention en milieu hospitalier

Présentée par
Sarah Bérubé

A été évaluée par un jury composé des personnes suivantes

Serge Lecours
Président-rapporteur

Serge Sultan
Directeur de recherche

France Dupuis
Membre du jury

Véronique Parent
Examinateur externe

Résumé

L’hémophilie est une maladie héréditaire incurable qui ralentit la coagulation sanguine. Le plan de traitement vise à prévenir les saignements pouvant se produire dans les articulations, puisque ceux-ci peuvent causer des dommages à long terme dans les structures internes. Le traitement consiste en l’administration régulière et continue d’injections de concentré de facteur de coagulation. Le plan de traitement inclut aussi des indications sur les activités physiques sécuritaires et celles à éviter pour les patients atteints de la maladie. Peu de recherches psychosociales ont été conduites en hémophilie à ce jour.

L’objectif de cette thèse, composée de 5 articles, était de déterminer les facteurs sociocognitifs susceptibles d’influencer l’adhésion aux recommandations chez les enfants et les adolescents atteints d’hémophilie et de proposer des pistes d’intervention. Pour ce faire, nous nous sommes appuyés sur trois modèles théoriques qui apportent, en complémentarité, un éclairage sur des aspects centraux de l’adhésion.

Dans le premier article, nous avons traduit en français un instrument de mesure de l’adhésion au traitement par injection et nous avons fourni les données psychométriques préliminaires de l’instrument. Les données ont appuyé son utilisation auprès des jeunes patients, ce qui nous a permis de l’utiliser dans la thèse.

Dans le deuxième article, nous avons utilisé la théorie du comportement planifié pour comprendre comment les jeunes se positionnent face aux recommandations de l’équipe médicale en ce qui a trait au traitement par injection et à l’activité physique. Pour cela, nous avons mesuré leur attitude, leur perception de contrôle comportemental, la norme sociale subjective et le comportement passé. Nous avons évalué si ces facteurs étaient liés à l’intention de suivre les recommandations. Les résultats ont indiqué que ces facteurs expliquaient une grande proportion de la variance de l’intention de suivre les recommandations sur l’activité physique, surtout pour celles dites « à risque ». Ayant constaté une bonne adhésion au traitement par injection chez les participants et une adhésion plus faible en ce qui a trait à l’activité physique, nous avons décidé de nous concentrer sur l’activité physique pour la suite.

Dans le troisième article, nous avons utilisé le modèle du sens commun pour comprendre comment les patients se représentent leur maladie et si ces représentations pouvaient avoir un impact sur leur pratique d’activé physique. Les perceptions étaient plus négatives pour les domaines suivants : les conséquences de la maladie, le nombre de symptômes et l’impact émotionnel. Lorsque nous avons classé les participants selon leur profil d’activité physique, le sous-groupe « adhésion faible », c’est-à-dire celui qui suivait moins et avait moins l’intention de suivre les recommandations, avait significativement plus de préoccupations par rapport à la maladie et percevait plus de symptômes en moyenne.

Le quatrième article souligne l’intérêt du modèle du soutien à l’autonomie de la théorie de l’autodétermination dans l’intervention en hémophilie et consiste en une adaptation pratique de ce modèle aux enjeux spécifiques de la maladie.

Le cinquième article porte sur un livret d’information que nous avons développé utilisant la théorie de l’autodétermination. Nous avons utilisé des méthodes quantitatives et qualitatives pour évaluer la réception du livret chez les parents et pour apporter des modifications à celui-ci.

Mots clefs : hémophilie, adhésion au traitement, activité physique, comportements de santé, théorie du comportement planifié, théorie de l’autodétermination, autorégulation, perceptions de la maladie, soutien à l’autonomie, psychologie de la santé, pédiatrie

Abstract

Hemophilia is an incurable bleeding disorder that slows the blood clotting process. The treatment plan aims to prevent bleeding episodes that occur in the joints, since repeated bleeding episodes can cause long-term damage to internal structures. The treatment plan consists of regular infusions of the missing clotting factor and specific recommendations as to which physical activities are safe and which are to be avoided.

The objective of this thesis, composed of 5 articles, was to determine the socio-cognitive factors likely to influence the adherence of children and adolescents to the recommendations of the treatment plan, and to suggest avenues for intervention. To do this, we relied on three theoretical models which, together, shed light on the central aspects of adherence: the perspective of young patients on the medical recommendations, their perceptions of the illness and, finally, avenues of intervention to promote better adherence in those young patients in the long-term.

In the first article, we translated to French an instrument that measured adherence to the infusion therapy, and we provided the preliminary psychometric data of the instrument. The data supported the use of the instrument with young patients, which allowed us to use it in the thesis.

In the second article, we used the theory of planned behavior to understand how young people perceive the recommendations concerning infusion treatment and physical activity. For this, we measured their attitude towards the behavior, their perception of behavioral control, the subjective social norm and past behavior. We assessed whether these factors were related to the intention to follow the recommendations. The results indicated that these factors explain a large proportion of the variance in intention to follow physical activity recommendations, especially for those at risk of causing bleeding. As adherence to injection therapy was high in our sample while adherence to physical activity recommendations was lower and more variable among participants, we decided to focus on the latter for the next articles.

In the third article, we wanted to understand how patients view their illness and whether illness perceptions could have an impact on their physical activity practice. For this we used the common-sense model. Perceptions were more negative for the following areas: the consequences of hemophilia, the number of symptoms and the emotional impact. When we ranked participants according to their physical activity profile, the subgroup that followed the recommendations less closely and had a lower intention of following the recommendations had significantly more concerns and perceived more symptoms.

The fourth article highlights the interest of the autonomy support model in hemophilia intervention and consists of a practical adaptation of this model to the specific challenges of the illness.

The fifth article is about the development of an information booklet that we created using the concepts of this theory. We used quantitative and qualitative methods to assess the parents' acceptability of the booklet and modified the booklet according to their suggestions.

Keywords : hemophilia, adherence to treatment, physical activity, health behaviors, theory of planned behavior, self-determination theory, self-regulation, perceptions of illness, autonomy support, health psychology, pediatrics

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Liste des abréviations

BRIEF-IPQ: Brief Illness Perception Questionnaire
FMH : Fédération mondiale de l'hémophilie
MSC : Modèle du sens commun (MSC)
PA : Physical activity
SCH : Société canadienne d'hémophile (SCH)
SDT: Self-determination theory
TCP : Théorie du comportement planifié (TCP)
TPB: Theory of planned behavior
VERITAS-Pro : Validated Hemophilia Regimen Treatment Adherence Scale--Prophylaxis
VERITAS-PRN: Validated Hemophilia Regimen Treatment Adherence Scale—On demand

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Introduction

L'hémophilie

L'hémophilie est une maladie héréditaire qui prolonge la durée normale de la coagulation sanguine. Elle est causée par une carence en un facteur de coagulation particulier, voire par une absence totale de celui-ci. Il s'agit du facteur VIII dans l'hémophilie dite de type A, et du facteur IX dans l'hémophilie dite de type B. L'hémophilie A est la forme la plus courante et sa prévalence est estimée à un peu moins de 1/10 000, ce qui représente environ 2 500 Canadiens. L'hémophilie B se produirait dans une proportion de 1/50 000, soit chez environ 600 Canadiens (Canadian Hemophilia Society, 2018). Une méta-analyse récente rapporte que la prévalence pourrait même être plus élevée, soit une estimation de près de 1.7/10 000 pour l'hémophilie A (Iorio, Stonebraker, Chambost, Makris, & Coffin, 2019). La majorité des personnes atteintes d'hémophilie sont des hommes.

On peut également décrire l'hémophilie en fonction de sa sévérité: légère (5 à 30 % du taux normal de facteur de la coagulation), modérée (1 à 5 % du taux normal) ou sévère (moins de 1% du taux normal) (Canadian Hemophilia Society, 2018; Iorio et al., 2019). Les patients atteints d'hémophilie font l'expérience de saignements internes et externes d'une durée prolongée par rapport à la normale, sans que les saignements soient plus rapides ou abondants (Canadian Hemophilia Society, 2018). Les patients atteints d'une forme sévère font face à ces épisodes de saignement prolongés plus fréquents par rapport à ceux affectés de la forme légère et modérée et il n'est pas rare, chez ceux-ci, que les saignements se produisent en l'absence de blessure avérée (saignements spontanés) (White et al., 2001). Contrairement aux saignements externes, les saignements qui se produisent à l'intérieur du corps sont particulièrement dangereux. Lorsqu'un saignement se produit dans un organe vital, comme le cerveau, il peut même être fatal (Payne et al., 2017). Les saignements dans les articulations, quant à eux, amènent un risque élevé de lésions pouvant mener, surtout s'ils sont répétés, à l'épaississement de la membrane synoviale, à des limites dans l'amplitude de mouvement et à de la douleur chronique (Bayer, 2016; Nazzaro, Owens, Hoots, & Larson, 2006). Les genoux, les coudes et les chevilles sont les endroits les plus couramment touchés (Nazzaro et al., 2006). La recherche a démontré que le respect des recommandations reliées au traitement par injections et à l'activité physique diminue de manière très importante les risques de complications à long terme (Philpott, Houghton, & Luke, 2010).

L’autosoin en hémophilie

La plupart du temps, le premier épisode de saignement se produit au cours de la première année de vie (Bullinger & von Mackensen, 2008). Bien que l’hémophilie demeure incurable encore à ce jour, elle peut être contrôlée par un traitement à vie qui consiste généralement en l’administration, par voie intraveineuse, de concentré de facteur VIII ou IX, selon certains horaires et dosages (Bullinger & von Mackensen, 2008). Le traitement par injection peut se faire à la maison pour la plupart des patients et est très efficace pour réduire le nombre de saignements (Panicker, Warrier, Thomas, & Lusher, 2003). Celui-ci peut s’effectuer sur demande, c’est-à-dire suite à une blessure ou lorsque le patient présente des symptômes annonçant un saignement, ou par traitement prophylactique, c’est-à-dire par un programme d’injections régulier et continu plusieurs fois par semaine qui vise à prévenir les saignements avant que ceux-ci se produisent. Les traitements sont très coûteux (Globe, Curtis, & Koerper, 2004). Au Canada, le coût du traitement prophylactique est remboursé par le régime public d’assurance-maladie. Bien que le traitement par injections soit utile pour diminuer le nombre de saignements, il demeure partiellement efficace. En effet, la dose injectée et la diminution graduelle de la concentration présente dans le sang une fois l’injection administrée ne permettent pas aux patients d’atteindre en tout temps un taux de concentration de facteur de coagulation dans le sang qui équivaut à celui d’une personne non atteinte de la maladie. D’autres mesures sont donc à prendre afin de prévenir les saignements.

Le plan de traitement inclut généralement des recommandations entourant la pratique d’activité physique. En plus des bénéfices connus du sport sur la santé physique et psychologique des enfants, d'autres, plus spécifiques à l’hémophilie, s’y ajoutent. En effet, développer des muscles robustes entourant les articulations, acquérir une bonne coordination dans le mouvement, ainsi que le maintien d’un poids équilibré réduit les risques de saignement et de complications (Buzzard, 1997; Nazzaro et al., 2006; Tiktinsky, Falk, Heim, & Martinovitz, 2002; Wearing, Hennig, Byrne, Steele, & Hills, 2006; Wittmeier & Mulder, 2007). La Fédération mondiale de l’hémophilie (FMH) mentionne dans ses lignes directrices que la décision concernant le sport le plus approprié pour chaque patient devrait se prendre en collaboration avec l’équipe de soin (Srivastava et al., 2013a). À cet effet, la FMH et la Société canadienne de l’hémophilie (SCH) soulignent que la plupart des activités physiques comportent une part de risque de saignement articulaire, mais que certains facteurs sont plus susceptibles d’aggraver le risque d’hémorragie et de blessures graves : le contact, la vitesse, la force de l’impact, la hauteur et l’imprévisibilité des conditions (Srivastava et al., 2013a; Zourikian, 2010). Plusieurs activités sportives généralement pratiquées par les enfants et les adolescents sont donc considérées comme dangereuses pour les personnes atteintes d'hémophilie, comme le soccer, le football et le hockey.

En hémophilie, l'autosoin peut donc être défini en référence à trois dimensions : (1) administrer le traitement par injections (2) pratiquer de l'activité physique recommandée et (3) éviter les sports déconseillés. Nous pouvons aussi ajouter à cela certaines mesures supplémentaires comme le fait de sécuriser l'environnement physique pour les enfants en bas âge et de s'assurer que des mesures sont prises lors de chirurgies ou d'extractions dentaires.

Les difficultés en lien avec l'autosoin

Chaque épisode de saignement rend les articulations plus fragiles et plus vulnérables à d'autres épisodes saignements. Considérant que deux ou trois saignements dans la même articulation peuvent causer des dommages irréparables et compromettre la qualité de vie, il est important de maximiser la prévention de ceux-ci (Gringeri, Ewenstein, & Reininger, 2014). Il a été démontré que la prophylaxie, lorsqu'elle est commencée en bas âge, prévient la détérioration des articulations (Nilsson, Berntorp, Lofqvist, & Pettersson, 1992). Cependant, la littérature soulève des enjeux qui doivent être pris en compte, particulièrement le fait qu'une bonne part des sujets atteints d'hémophilie développent encore des complications de la maladie, souvent en lien avec une mauvaise adhésion au traitement (Kraft et al., 2012). Au Canada, une étude a mis en lumière que 15% des enfants et 27% des jeunes adultes atteints d'hémophilie qui participaient à l'étude avaient une « articulation cible », c'est-à-dire, une articulation dans laquelle s'étaient produit au moins trois saignements en trois mois (St-Louis et al., 2016). Il se produit à l'intérieur d'une articulation cible des mécanismes mécaniques, chimiques et enzymatiques qui risquent fortement de causer des dommages à long terme.

Dans la suite de la thèse, le terme adhésion se réfère à la définition de l'Organisation mondiale de la santé , soit « le degré avec lequel le comportement de la personne - prendre la médication, suivre le régime et/ou exécuter les changements dans le style de vie - correspond aux recommandations ayant été convenues avec un professionnel de la santé » [traduction libre] (World Health Organization, 2003).

L'adhésion au traitement par injection diminue considérablement pendant l'adolescence. Dans un sondage mondial effectué dans 147 centres de traitements, Geraghty et ses collègues ont observé que l'adhésion au traitement prophylactique était plus élevée chez les enfants jusqu'à l’âge de 12 ans avec 90 % d’adhésion « élevée » ou « très élevée » (Geraghty et al., 2006a). Toutefois, le pourcentage chutait à 54 % chez les adolescents de 13 à 18 ans. Cette période coïncide avec le passage vers l’autoadministration des injections. En effet, c’est généralement dans cette tranche d’âge que les jeunes apprennent graduellement à administrer eux-mêmes leurs injections plutôt que ce soient leurs parents qui en soient entièrement responsables. Dans une étude américaine, moins du tiers des adolescents rapportaient traiter leurs saignements en moins d’une heure, comme recommandé (Nazzaro et al., 2006). Au Canada, un sondage a démontré que seulement 50% des médecins pensaient que le taux d’adhésion de leurs patients pédiatriques se situait entre 80 et 100% (Chan, Decker, & Warner, 2011). Ces études mettent en lumière qu’une bonne partie des patients pédiatriques ne prennent pas leurs injections comme recommandé et s’exposent ainsi à un risque accru d’épisodes de saignements. Ces épisodes de saignement peuvent provoquer de la détresse, obliger des périodes de repos et d’alitement, et mener à des hospitalisations et au développement de problèmes articulaires chroniques (Cassis, 2007).

Des enjeux similaires concernent les recommandations en ce qui a trait à l’activité physique des jeunes atteints d’hémophilie. En effet, alors que l’exercice physique est fortement recommandé à condition de prendre les précautions nécessaires, une étude américaine rapportait que 60 % des jeunes atteints d’hémophilie gèrent leur maladie en évitant les activités physiques (Nazzaro et al., 2006). Les enfants atteints d’hémophilie percevraient aussi plus de barrières à l’activité physique et verraient moins l’activité physique comme une alternative positive par rapport à la sédentarité que les sujets en santé (Buxbaum, Ponce, Saidi, & Michaels, 2010). Il est aussi possible que les jeunes se sentent moins compétents ou ne comprennent pas bien les bénéfices de l’activité physique sur leur condition de santé. Il a été démontré que le développement de compétences spécifiques à l’activité physique, comme la coordination motrice, est un important prédicteur de l’activité physique future (Lopes, Rodrigues, Maia, & Malina, 2011). En bougeant, les jeunes développent leurs habiletés physiques et ils sont plus enclins à s’engager dans des activités physiques par la suite. Il est donc important de favoriser leur développement à ce niveau, malgré les limitations inhérentes à leur condition de santé. À l’opposé de la sédentarité, une grande proportion des jeunes atteints d’hémophilie pratiquent des activités dites « à grand impact » comme le football, le basketball et le soccer (Ross, Goldenberg, Hund, & Manco-Johnson, 2009). Ces activités sont généralement déconseillées étant donné qu’elles comportent un risque important de causer des saignements (Tiktinsky et al., 2009). Ces résultats sont préoccupants, d’autant plus si nous considérons que l’activité physique de l’enfant et de l'adolescent est un important prédicteur de l'activité physique à l'âge adulte (Perkins, Jacobs, Barber, & Eccles, 2004).

Il est intéressant de se demander ce qui pousse certains jeunes à être impliqués et motivés pour leur plan de traitement et ce qui, au contraire, en pousse d'autres à ne pas suivre les recommandations, malgré les conséquences connues sur leur état de santé. Nous savons que les enfants et les adolescents tendent à mettre l'accent sur le présent et peuvent avoir de la difficulté à prévoir les conséquences à long terme de leur comportement (Petrini & Seuser, 2009). L'enfance et l'adolescence constituent aussi des périodes de vie marquées par des questionnements et des préoccupations concernant l'identité, l'estime de soi et les relations interpersonnelles. Dans le cas des adolescents atteints d'hémophilie, pratiquement tous les aspects de la vie peuvent en être affectés: les aspirations professionnelles, les activités sportives, les voyages et les rencontres amoureuses, etc. (Nazzaro et al., 2006). La période de l'adolescence amène donc les parents et les jeunes patients atteints d'hémophilie à vivre des inquiétudes majeures (Geerts, van de Wiel, & Tamminga, 2008). Les adolescents sont souvent préoccupés par le fait d'être jugés par leurs pairs en raison des limitations de la maladie (Geraghty et al., 2006a). La diminution de l'adhésion au traitement à l'adolescence chez les patients atteints de maladies chroniques est bien documentée dans la littérature (Taddeo, Egedy, Frappier, 2008). Plusieurs auteurs soulignent que le processus de préparation à la transition vers l'âge adulte devrait commencer bien avant l'adolescence et s'opérer de manière graduelle afin de favoriser une meilleure continuité dans l'adhésion (Reiss, Gibson, Walker, 2005 ; Wong et al., 2010). Bien que tous les auteurs soulignent la vulnérabilité des patients pendant cette période de transition, très peu de travaux portent spécifiquement sur la préparation du passage du soin administré par les parents au soin dit autonome (Breakey, Blanchette, & Bolton-Maggs, 2010; Geerts et al., 2008).

Études sur l'adhésion au traitement en hémophilie

La plupart des études cherchant à identifier les facteurs influençant l'adhésion au traitement par injection chez les jeunes atteints d'hémophilie procèdent sous forme de sondage, en demandant aux patients, aux parents et aux professionnels de la santé d'identifier les raisons qui expliquent les difficultés d'adhésion (Geraghty et al., 2006; Hacker, Geraghty, & Manco-Johnson, 2001; Nazzaro et al., 2006; Schrijvers, Uitslager, Schuurmans, & Fischer, 2013). Les raisons de non-adhésion rapportées dans ces études sont la disparition ou la réduction des symptômes, un accès veineux difficile, l'oubli, le manque de temps, le fait de ne pas reconnaître les signes de saignements, le fait de ne pas comprendre les bénéfices du traitement, le déni de la maladie, la rébellion adolescente, le manque d'implication parentale et l'interférence avec le style de vie (De Moerloose, Urbancik, Van Den Berg, & Richards, 2008; Geraghty et al., 2006b; Hacker et al., 2001; Nazzaro et al., 2006). Ces sondages ne calculent cependant pas les corrélations entre les facteurs proposés et une mesure de l'adhésion, ce qui ne nous permet pas d'apprécier si ces facteurs ont un impact réel sur le comportement. De plus, ces études n'utilisent pas un modèle théorique pour expliquer l'adhésion au traitement. Or, les modèles théoriques servent de terrains communs pour les chercheurs, facilitent la reproductibilité de la recherche et le développement d'instruments de mesure valides (Lippke & Ziegelmann, 2008). Comme les modèles théoriques présentent aussi les liens qui unissent les différents prédicteurs du comportement et sont parcimonieux quant au nombre de facteurs impliqués, ils facilitent la conception et l'évaluation des interventions et donc l'applicabilité clinique de la recherche. Dans une revue de la littérature récente sur les facteurs reliés à l'adhésion au traitement en hémophilie (enfants et adultes confondus), près du tiers des études portait sur des facteurs non modifiables, comme les données sociodémographiques (Sultan, El-Hourani, Rondeau, & Garnier, 2018). Les auteurs constataient également que les facteurs sociaux liés à l'adhésion n'ont pratiquement pas été étudiés en hémophilie (Sultan, El-Hourani, Rondeau, & Garnier, 2018).

En comparant les données disponibles sur les prédicteurs de l'adhésion au traitement en hémophilie avec les données disponibles sur le diabète, une maladie qui requiert elle aussi un traitement par injections, nous constatons des lacunes importantes en hémophilie. La littérature sur le diabète est riche en ce qui a trait aux facteurs associés à l'adhésion au traitement, ce qui permet d'établir une classification des prédicteurs de l'adhésion. Cette classification aide à identifier les domaines sur lesquels les efforts d'intervention devraient porter (Davies et al., 2013 ; Polonsky, & Henry, 2016; Sultan, El-Hourani, Rondeau, & Garnier, 2018). En ce qui a trait au diabète, il est clairement démontré que les facteurs reliés à l'environnement social, comme la qualité de la relation parent-enfant, ainsi que des facteurs reliés à l'équipe de soin, comme la relation soignant-patient ou l'accès à un programme de transition, ont un impact significatif sur l'adhésion au traitement. Il n'est d'ailleurs pas surprenant que pour cette maladie, plusieurs interventions visant à favoriser un meilleur contrôle glycémique chez les jeunes patients aient déjà été testées empiriquement (Hood, Rohan, Peterson, & Drotar, 2010). Bien que nous puissions nous inspirer des modèles testés dans le domaine du diabète, il demeure important d'étudier les enjeux spécifiques à l'hémophilie, puisque ceux-ci diffèrent grandement de ceux retrouvés chez les enfants atteints de diabète. Notamment, les enfants atteints d'hémophilie n'ont pas de système de monitorage équivalent au contrôle glycémique. Ainsi, ils n'ont pas d'indicateur leur permettant de savoir si leur autosoin est optimal, outre l'absence d'épisodes de saignement.

Tel que nous l'avons mentionné précédemment, en plus de l'adhésion au traitement par injection, l'activité physique est au centre du plan de traitement en hémophilie. La littérature portant sur l'étude des déterminants de l'activité physique est riche en ce qui concerne les enfants et les adolescents en santé (Sallis, Prochaska, & Taylor, 2000; St-Louis et al., 2016). Celle-ci soulève que des facteurs socioéconomiques, personnels et environnementaux, comme l'autoefficacité et le sentiment de contrôle, la perception de barrières et de bénéfices à l'activité physique, le support parental et le « modeling » des parents et des pairs sont associés à l'activité physique chez les enfants et chez les adolescents. Cependant, les études sur une population en santé, bien qu'utiles pour le développement d'interventions en santé publique, ne peuvent pas nécessairement être transférées aussi facilement au contexte de la maladie chronique. En hémophilie par exemple, la maladie a un impact sur le choix possible d'activités, et un non-respect des recommandations peut avoir des conséquences directes et néfastes sur la santé à court, moyen et long terme. De plus, l'activité physique fait souvent l'objet de discussions et d'interactions entre le personnel soignant, les parents et les patients, ce qui soulève des enjeux particuliers qui doivent être pris en compte dans les études afin de soutenir l'intervention. Dans d'autres maladies, certains facteurs sont ressortis comme ayant une importance majeure sur la pratique d'activité physique des patients. Chez les enfants et adolescents atteints d'asthme par exemple, une revue de la littérature a démontré qu'une mauvaise connaissance des symptômes pouvait affecter la pratique d'activité physique (Williams, Powell, Hoskins, & Neville, 2008). Peu d'études existent sur les déterminants psychosociaux de l'activité physique chez les patients atteints d'hémophilie. De plus, les études disponibles lient peu souvent les facteurs étudiés à une mesure du niveau d'activité physique et ne distinguent pratiquement jamais les activités physiques recommandées des activités déconseillées en n'étudiant que le niveau global d'activité physique. Cela est problématique puisque les recommandations et les impacts sont différents s'il s'agit d'une activité recommandée ou d'une activité à risque de causer des saignements.

Récemment, deux questionnaires (VERITAS-Pro, VERITAS-PRN) évaluant de manière standardisée l'adhésion à la prophylaxie et au traitement sur demande ont été développés, ce qui représente une avancée importante dans l'étude de l'adhésion au traitement en hémophilie (

Duncan, Kronenberger, Roberson, & Shapiro, 2010; Duncan, Kronenberger, Roberson, & Shapiro, 2010). Comprendre les déterminants de l'adhésion aux recommandations permettrait d'outiller les professionnels de la santé afin de favoriser une meilleure adhésion chez leurs jeunes patients. À ce jour, le moyen le plus utilisé par ceux-ci pour favoriser l'adhésion est l'éducation à propos de l'hémophilie (Chan et al., 2011). Ce mode d'intervention présente des limites, puisqu'une meilleure compréhension de l'hémophilie ne garantit pas nécessairement un autosoin adéquat. Par exemple, lors d'une étude, 41 % des patients ont déjà décidé de ne pas suivre un traitement prophylactique prescrit, malgré un haut niveau de connaissance de l'hémophilie (Geraghty et al., 2006a).

Approche théorique sur l'adhésion au traitement

Dans les années 70, nous avons assisté à un désenchantement face à un modèle de conceptualisation de la santé et des maladies strictement biomédical, qui voit le corps comme étant séparé des processus psychologiques et sociaux. Cela a mené au développement du modèle biopsychosocial, plus adapté à la compréhension des maladies chroniques, dont la prévalence a augmenté en parallèle avec l'espérance de vie (Lyons & Chamberlain, 2006). Le modèle biopsychosocial s'intéresse non seulement aux facteurs biologiques reliés à la santé et à la maladie, mais également aux facteurs psychologiques, sociaux et comportementaux. Cette perspective a amené plusieurs disciplines à s'intéresser aux enjeux de santé physique comme la sociologie et l'anthropologie (Lyons & Chamberlain, 2006). De cet engouement est née la psychologie de la santé, dont un des axes centraux est la prédiction des comportements de santé (Conner & Norman, 2005; Sarafino & Smith, 2014).

À ce jour, dans d'autres maladies que l'hémophilie, l'autosoin a été l'objet de modélisations faisant appel aux modèles théoriques de prédiction du comportement comme la théorie du comportement planifié (TCP) (*theory of planned behavior*) de Azjen et le modèle du sens commun de Leventhal (*common-sense model*) (Ajzen, 1991; Diefenbach & Leventhal, 1996; Jones, Smith, & Llewellyn, 2016; Lippke & Ziegelmann, 2008; Rich, Brandes, Mullan, & Hagger, 2015).

Lacunes des études antérieures

En résumé, l'importance d'un bon autosoin (incluant l'adhésion au traitement et le suivi des recommandations concernant l'activité physique) et ce, dès le plus jeune âge, a été largement démontrée dans les études antérieures. Ce niveau d'autosoin doit persister lorsque l'enfant devient plus autonome au cours de l'adolescence. Il est important d'étudier les facteurs qui poussent un enfant à suivre ou à ne pas suivre les recommandations, puisque ce sont ces facteurs qui pourront par la suite être ciblés par l'équipe médicale dans leurs interventions afin de permettre au jeune patient d'être engagé et motivé dans le traitement de sa maladie. À ce jour, plusieurs études portent sur l'adhésion au traitement dans d'autres maladies chroniques et des interventions efficaces ont été développées. En contraste, la littérature psychosociale en hémophilie est très mince dans sa compréhension de l'adhésion au traitement et des facteurs susceptibles d'avoir un impact sur les comportements. Les particularités de l'hémophilie, dont l'importance de reconnaître et de traiter rapidement les saignements, l'invisibilité de la maladie en l'absence de symptômes et les limitations importantes dans le domaine de l'activité physique, font d'elle une condition de santé aux enjeux uniques. Les recommandations en hémophilie peuvent être particulièrement difficiles à suivre pour les jeunes patients. Comme nous l'avons vu, des études sont disponibles sur les niveaux d'adhésion, mais peu d'études se concentrent sur les déterminants psychosociaux de l'adhésion. Pratiquement aucune donnée psychosociale n'est disponible concernant les facteurs influençant l'activité physique chez les jeunes atteints d'hémophilie. Cela fait en sorte que les professionnels de la santé peuvent manquer d'information ou de matériel pour intervenir efficacement avec leurs patients. De plus, les parents n'ont pas nécessairement accès à de l'information validée scientifiquement pour les guider dans leur façon de communiquer à propos de la maladie avec leurs enfants, comme ce peut être le cas chez les enfants atteints de cancer par exemple.

Orientation du programme de thèse et objectifs généraux

Nous avons développé un programme de recherche afin de mieux comprendre le phénomène de l'adhésion au traitement chez les jeunes atteints d'hémophilie, incluant l'adhésion aux recommandations sur l'activité physique.

Dans un premier temps, nous désirions décrire la qualité de l'adhésion chez les jeunes atteints de cette condition. Dans l'article 1, nous avons commencé par traduire un instrument de mesure de l'adhésion au traitement déjà existant. Cela nous a aussi permis de dresser un portrait détaillé de l’adhésion au traitement par injection dans notre population. Suite à cet article, nous ne savions toujours pas si les enjeux d’adhésion et de motivation étaient plus prononcés en ce qui a trait au traitement par injection ou en ce qui concerne l’activité physique. L’article 2 visait à répondre à cette question en comparant la motivation des jeunes à suivre les recommandations pour leur plan de traitement et leur motivation à suivre les directives entourant l’activité physique.

En deuxième temps, dans l’article 2 et 3, nous avons souhaité investiguer les facteurs pouvant expliquer la motivation des jeunes patients pour leur plan de traitement. Pour ce faire, nous avons fait appel à des modèles théoriques connus, soit la théorie du comportement planifié, dans l’article 2, et le modèle du sens commun, dans l’article 3. Nous décrirons ces deux modèles dans la suite du document. Il faut savoir qu’en avançant dans le programme de thèse et suite à l’obtention des résultats des deux premiers articles, nous avons pris la décision de nous concentrer seulement sur les déterminants de l’activité physique pour la suite de la thèse. En effet, les résultats ont mis en lumière que l’adhésion au traitement par injection, ainsi que l’intention de suivre les recommandations sur la prophylaxie et le traitement sur demande étaient très élevées chez les jeunes patients (créant ainsi un effet plafond). En revanche, leur intention de pratiquer les activités physiques recommandées et déconseillées était plus variable selon les participants. De plus, l’activité physique est rarement étudiée d’un point de vue psychosocial en hémophilie, ce qui a alimenté notre curiosité envers cet enjeu. Une des particularités de notre projet de recherche est que nous distinguons deux types d’activité physique, soit celle à risque et celle qui est conseillée par les professionnels de la santé, plutôt que d’utiliser un score global d’activité physique comme ce qui est fait dans le peu d’études psychosociales à ce sujet. En somme, l’article 2 et 3 ont permis d’identifier les différences entre les jeunes qui adhèrent aux recommandations d’activité physique et ceux qui n’y adhèrent que partiellement. Nous avons pu identifier des facteurs cibles modifiables qui pourraient faire l’objet d’intervention.

Finalement, dans un dernier temps, nous nous sommes questionnés à savoir quelle serait la meilleure façon d’intervenir sur ces facteurs cibles. Cette réflexion nous a amenés à constater qu’il existait peu de lignes directrices dans la littérature sur la façon dont nous pouvons modifier ces facteurs, même si ceux-ci sont issus de modèles théoriques bien étudiés. Il y a pourtant consensus dans la littérature sur le fait que les interventions psychosociales devraient être ancrées dans un modèle théorique et s’appuyer sur les données probantes (Araújo-Soares, Hankonen, Presseau, Rodrigues, & Sniehotta, 2018). Ne trouvant pas dans les théories utilisées dans les articles 2 et 3 assez d’éléments pour conceptualiser une intervention, nous avons dû chercher ailleurs pour trouver réponse à nos questions. Nous avons alors fait appel à un troisième courant théorique, soit la théorie de l’autodétermination. Cette théorie est complémentaire à la théorie du comportement planifié et au modèle du sens commun, puisqu’elle donne des indications précises sur la façon de conceptualiser l’intervention, et ce, peu importe le modèle théorique duquel sont tirés les facteurs. Nous reviendrons sur ce point plus en détail dans la discussion générale de la thèse.

Cette thèse rend compte d’une démarche exploratoire. Nous visions à établir certaines bases de compréhension sur les enjeux entourant l’adhésion et la motivation des jeunes pour leur plan de traitement. L’utilisation de plusieurs modèles théoriques est en ce sens pertinente puisque la littérature psychosociale dans le domaine l’hémophilie est très peu développée. Aucun modèle ne semble à ce jour pleinement satisfaisant ou complet pour étudier les comportements des patients en psychologie de la santé (Lippke & Ziegelmann, 2008). Dans la littérature actuelle, nous constatons que des efforts sont faits pour combiner les différents modèles. À ce jour, ces efforts aboutissent à des modèles très complexes qui font l’objet de critiques concernant leur caractère non parcimonieux et leur faible transférabilité clinique (Lippke & Ziegelmann, 2008; Prochaska, Wright, & Velicer, 2008). Afin de clarifier et de justifier l’orientation de chacune des cinq études incluses dans cette thèse, nous présentons une introduction avant chaque article.

Introduction à l'article 1

L'objectif premier de cet article visait à traduire en français le VERITAS-Pro, mesurant l'adhésion au traitement prophylactique en hémophilie, et le VERITAS-PRN, mesurant, quant à lui, l'adhésion au traitement sur demande (Duncan et al., 2010a, Duncan et al., 2010b). Nous souhaitions vérifier l'applicabilité du questionnaire auprès d'une population francophone québécoise et décrire sommairement la structure de cet outil, en examinant la consistance interne des mesures qui en découlent. C'est ce questionnaire qui a permis d'évaluer l'adhésion au traitement par injection dans la suite de la thèse. Cette traduction sera aussi désormais disponible aux autres chercheurs qui désireraient l'utiliser afin d'étudier l'adhésion dans une population francophone.

Dans cet article, nous avons également décrit l'adhésion au traitement des jeunes, telle que rapportée à cet instrument par les parents et par les jeunes patients, et ce, pour différentes conditions. Par exemple, nous avons pu comparer les niveaux d'adhésion rapportés lorsque ce sont les parents qui administrent les injections à ceux rapportés lorsque ce sont les jeunes patients qui en sont eux-mêmes responsables. Ces résultats nous ont aussi permis de déterminer les domaines d'adhésion les plus problématiques. En effet, les deux questionnaires contiennent plusieurs échelles qui représentent différents aspects de l'adhésion, comme la capacité à se rappeler de faire les injections et l'habitude de bien planifier l'approvisionnement en concentré de facteur.

Article 1

Measuring adherence in pediatric hemophilia patients: French-language adaptation of the VERITAS-Pro and VERITAS-PRN

Sarah Bérubé PhD(c) [1,2], Émélie Rondeau[1], Serge Sultan PhD[1,2,3]

[1] CHU Sainte-Justine, Montreal, Quebec, Canada
[2] Departement of Psychology, Université de Montréal, Montréal, Québec, Canada
[3] Department of Pediatrics, Université de Montréal, Montréal, Québec, Canada

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Abstract

VERITAS-Pro and VERITAS-PRN are instruments to assess adherence to treatment of people with hemophilia. Low adherence can limit treatment effectiveness and result in increased bleeding episodes and chronic pain. The purpose of this study was to adapt both questionnaires into French, report preliminary psychometric data in a group of pediatric patients and their parents, and compare ratings between young patients and their parents. The translation process followed these steps: (1) forward translation (2) backward translation (3) final version, and (4) testing. Translators were familiar with the pediatric community. Thirteen pediatric patients and twenty parents participated in the study. The translation process was conducted without major difficulties and children easily understood the French versions. Preliminary psychometric analyses showed good to excellent consistency for the total scale and most subscales. However, the consistency of the Dose (VERITAS-Pro) and the Plan (VERITAS-PRN) subscales were low. When pediatric patients were primary infusers, they reported more skipping of prophylaxis than in parents' estimation of their behavior. For episodic treatment, when parents were primary infusers, they reported better adherence for the Time subscale vs. children's estimation of their behavior. Parents also rated higher adherence on prophylaxis when they were themselves in charge of performing the infusions than when young patients were autonomous. Preliminary psychometric analyses support the use of the French versions for the total scale and most subscales. However, the results did not support the use of subscales Dose (VERITAS-Pro) and Plan (VERITAS-PRN) in the adapted versions. Further studies should be performed on larger samples.

Keywords : Hemophilia, Pediatrics, Adherence, Hematology, Translation

Introduction

Hemophilia is a rare and inherited bleeding disorder caused by a deficiency in a coagulation factor, most often factor VIII (hemophilia A) or factor IX (hemophilia B). The severe form is characterized by spontaneous bleeding, predominantly in joints and muscles. Over time, repeated bleeding can lead to arthropathy, chronic pain, and impaired health-related quality of life (A. Gringeri et al., 2014; Valentino, 2010). Joint bleeding can be greatly prevented with replacement therapy, which consists of intravenous infusions of the missing factor on a regular schedule (prophylaxis) or in case of injury or high risk for bleeding (episodic treatment) (Manco-Johnson et al., 2007).

In a recent literature review on adherence in hemophilia, nonadherence to prescribed prophylactic regimens was linked to a wide range of patients, condition and treatment-related factors, as well as healthcare system and socioeconomic factors (Thornburg & Duncan, 2017). Nonadherence was also shown to limit treatment effectiveness and resulted in poorer outcomes such as increased bleeding episodes and chronic pain (Thornburg & Duncan, 2017). In pediatric patients, poorer adherence can also result in a greater number of missed days of school (Krishnan, Vietri, Furlan, & Duncan, 2015). To prevent long-term damages in the joints, it is recommended that one should start prophylaxis early in life (Coppola, Tagliaferri, Di Capua, & Franchini, 2012).

Maintaining a rigorous treatment schedule can be challenging for patients, especially during adolescence, when patients learn to self-administer their treatment instead of their parents or caretakers doing so (Schrijvers et al., 2016). During the teenage years, adherence to treatment tends to decline drastically in hemophilia, as in other chronic illnesses (Geraghty et al., 2006b; Taddeo, Egedy, & Frappier, 2008b). The development of a reliable tool to assess adherence in hemophilia is crucial to the study of adherence, its determinant and its impact, especially in the more vulnerable populations such as adolescents. Accurately measuring adherence may usefully open on interventions designed to optimize adherence levels in order to improve bleeding prevention among this population.

The Validated Hemophilia Regimen Treatment Adherence Scale was developed to address this need (Duncan et al., 2010a; Duncan et al., 2010b). This standardized measure of adherence has two different versions to assess both adherence to prophylactic (VERITAS-Pro) and episodic treatment (VERITAS-PRN). Adaptations are now available in Spanish, Dutch and

Brazilian Portuguese (Cuesta-Barriuso et al., 2017; Ferreira, Leite, & Duncan, 2017; Lock et al., 2014).

To date, no French-language version is available for the French-speaking patients and families. The objectives of the current study were to translate and adapt the VERITAS-Pro and VERITAS-PRN from English into French, to report preliminary psychometric data of the adapted versions, and to compare ratings between children and their parents.

Methods

Participants

Participants were recruited at the Sainte-Justine UHC Hemostasis Center. All children with severe hemophilia were approached if they had an appointment during the recruitment period and met the inclusion criteria: 6-18 years of age, French speaking, on prophylaxis and/or episodic treatment. We excluded young patients who were diagnosed with a psychiatric disorder or had received their diagnosis less than a year before the study and were still adapting to the illness, in order to avoid outliers and collect answers from a relatively homogenous sample representative of the majority of young people with hemophilia. Parents were also recruited as long as they spoke French. Written informed consent was obtained from all participants. The Sainte-Justine UHC Research Ethics Board approved this project.

In total, twenty pediatric patients, aged 6-18 years old ($M = 12 \pm 4$) with hemophilia A or B were recruited. Of those, thirteen were able to complete the questionnaires (nine primary infusers and four whose parents were in charge of their treatment). The other ones were too young to be aware of their parent’s behaviors regarding infusion management and adherence. All participants were on prophylaxis in addition to episodic treatment in case of bleeding. The parents (thirteen mothers, seven fathers) completed the questionnaires. Demographic characteristics for the sample are displayed on Table 1.

VERITAS-Pro and VERITAS-PRN

The 24-item questionnaires include six subscales (VERITAS-Pro: Time, Dose, Plan, Remember, Skip, Communicate; VERITAS-PRN: Treat, Time, Dose, Plan, Remember, Communicate). Time refers to taking the treatment as schedule, Dose refers to infusing the prescribed dose, Plan refers to the organization around the supplies, Remember refers to the missed infusions, Skip deals with the doses purposely not administered, Communicate is for calling the treatment center when needed, and Treat makes reference to administering infusions when symptoms of bleeding occur. The responses to each item are scored on a 5-point Likert scale, ranging from “Always” to “Never”. Specifications on the meaning of each anchor point are given in the instructions. For example, "Always" means "100% of the time", while “Often” means “at least 75% of the time”. In addition, the instructions mention that the respondent must respond based upon their behavior in the last 3 months. Total scores range from 24 to 120, and subscale scores range from 4 to 20 with lower scores indicating higher adherence. Both questionnaires have shown adequate psychometric properties in US samples. Internal consistency reliability for the total VERITAS-Pro score and all subscales was good–to–excellent (Cronbach’s alphas between .67 and .94), and test–retest reliability correlations were very strong (Duncan et al., 2010a). For the VERITAS-PRN, Cronbach’s alphas for the total score and all subscales were greater than .66, except for the plan subscale ($\alpha = .48$) (Duncan et al., 2010b). Scores on the VERITAS-Pro and VERITAS-PRN have correlated moderately-to-strongly with global adherence rating by primary infusers and medical staff (Duncan et al., 2010a; Duncan et al., 2010b). Scores on the VERITAS-Pro correlated as well as with the percentage of recommended infusions administered from a web-based self-report log system and validated against pharmacy dispensation (Duncan et al., 2010a).

Translation and adaptation process

Three bilingual translators familiar with the pediatric community participated in the study: two whose mother tongue was French (JSL and SB, see acknowledgments) and one whose mother tongue was English (LG, see acknowledgments). The methodology used to translate and adapt the questionnaires was based on the *World Health Organization (WHO) Process of translation and adaptation of instruments* (World Health Organization, 2009).

The first step was a forward translation by two independent translators to produce two conceptually equivalent French versions of each the VERITAS-Pro and VERITAS-PRN. The focus was on cross-cultural and conceptual, rather than on linguistic/literal equivalence. The instruments needed to be easily understood by pediatric patients. Technical terms were avoided. The forward translations were then compared and discrepancies were discussed between translators until a consensus emerged to produce the intermediate French versions.

Using the same approach as that outlined in the forward translation, the intermediate French versions were then back translated into English by an independent translator. The translated questionnaires were then compared to the original English version. Differences were discussed to explore if important cross-cultural or conceptual aspect were involved. Modifications were made to achieve a pre-final French version of each questionnaire.

We also tested the pre-final versions by asking the first 5 children to give their general impressions of the questionnaires. They were asked if the items, instructions, and response scale were clear, easy to understand, and if there were any words they found difficult to comprehend, in line with the practice of cognitive interviews (Hak, van der Veer, & Jansen, 2008). All recruited children and parents were asked to complete the questionnaires and to think of the specific treatment regimen assessed by the questionnaire when filling them. Participants were assigned to two different groups depending on whether the child or the parent was the primary infuser. Children and parents who were not the primary infusers did also complete the questionnaires for comparison purpose.

Psychometric and statistical analysis

Descriptive statistics (mean scores, standard deviations) were calculated for each subscale. Lower scores represent higher adherence. Comparison between raters' scores we made using Wilcoxon rank tests. For this, we paired the child and the parent's estimate of adherence, ie their score on the VERITAS-Pro or -PRN questionnaires. We used Mann-Whitney U tests to compare parents' scores for each of the two categories (children as primary infusers vs. parents as primary infusers), and children's scores for each of the two categories. Mean scores in the present study were also compared to the validation study using Cohen's d where the difference is characterized as small ($d = 0.20$), medium ($d = 0.50$), or large ($d = 0.80$) (Cohen, 1992). Because of the small number of items in each subscale which might bias the alpha consistency coefficient, consistency was documented using inter-item correlations with values between .15 and .50 being considered acceptable (Clark & Watson, 1995; Streiner, 2003). For information purposes, Cronbach's alpha levels (α) are given as supplementary material. Item-total and item-subscale correlations were calculated, with a value above 0.30 to be considered as acceptable (Nunnally & Bernstein, 1994). As recommended, correlations were calculated by taking into account totals without the test item. For information purposes, we calculated the Pearson's correlation (r) between adherence to treatment and the number of bleeding episodes in the last year as informed by the nurse. All statistics were performed using SPSS software (SPSS Inc., Chicago, IL).

Results

Translators panel findings

The forward translation was conducted without major difficulties. The two versions of each questionnaire were very similar. Two significant issues were raised. The word *supplies* (items 10, 11, and 12, VERITAS-Pro) was translated to *équipement* [equipment] by translator 1 and to *fournitures* [supply] by translator 2. The concern was based on the fact that *fournitures* [supply] would better preserve the meaning of the English word but was thought to be harder to understand for children. Translators agreed to choose *fournitures* [supply] and to check for the understanding of the word during testing. Second, the word *prescribed* (item 15, VERITAS-Pro) was translated to *prescrit* [prescribed] by translator 1 and to *fourni* [provided] by translator 2. *Prescrit* [prescribed] was chosen because *fourni* [provided] would refer more to the idea of a written schedule.

After the back-translation process, minor discrepancies were encountered (eg timing vs. calendar; inconvenient vs. bothers me; occur vs. happen; convenient vs. practical moment). All of those were discussed until a consensus emerged. Translators were unsure of how to translate *I keep close track* (item 10, VERITAS-Pro). Two expressions were proposed to the first five participants: 1) *je suis de près* [I follow closely] and 2) *je fais un suivi rigoureux* [I follow rigorously]. Translators expressed some doubts on how to translate the word *convenient* (item 6, VERITAS-PRN). They agreed to use *convenable* [suitable] and to check the understanding of the word during testing.

The first five participants (*M* = 14 ± 3 yrs) were interviewed. All participants mentioned that the questionnaires were clear, easy to understand, and easy to answer. For items 10 to 12 (VERITAS-Pro), all children reported understanding the word *fournitures* [supply], and therefore, the wording was maintained. For item 10, patients had the choice between 1) *je suis de près* [I follow closely] and 2) *je fais un suivi rigoureux* [I follow rigorously]. None of the children were sure of the meaning of *rigoureux* [rigorously]. Thus, the first option was chosen. For item 6 (VERITAS-PRN), none of the children were certain what *convenable* [suitable] meant. Some children suggested to use *pratique* [practical] instead of *convenable* [suitable]. Thus, *pratique* was chosen. Finally, the word *colonoscopie* [colonoscopy] raised some questions from the children.

Preliminary psychometric data and Scores

Internal consistency

When examining inter-item correlations, internal consistency for VERITAS-Pro total scale and for most subscales was acceptable, except for the subscale Dose (total sample) and Plan (parent report only) for which correlations were low (see Table 2). For the Remember and Skip subscales, inter-item correlations were high (> .50) in the total sample reflecting items were very similar to each other and that there might be some redundancy. For VERITAS-PRN, inter-item correlations were acceptable for total scale and for most subscales (see Table 3). The Plan subscale was problematic with a very low inter-item correlation. The inter-item correlation in total sample was high for the Remember subscale pointing at some redundancy.

These results were consistent with the alpha analysis except for Dose and Communicate domains of the VERITAS-PRN for which the alpha coefficient for the parent report was low (.41 and .37) but had an acceptable inter-item correlation (.30 and .15). For problematic scales, discarding individual items did not manage to increase alphas to an acceptable level. Alphas indicated an excellent internal consistency for total scale.

Item analysis

All items were analyzed for item-total and item-subscale correlations (see supplementary file).

For VERITAS-Pro, item 14 from the Remember subscale (remember to infuse) was the one that contributed most to the total score with an item-total correlation of .78 for the total sample. For the total sample, none of the items of the Dose subscale correlated substantially with either their own subscale or to the total scale. It was also the case for item 9 of the Plan subscale (parent report only) and item 24 of the Communicate subscale.

In VERITAS-PRN, the most correlated item to the total score was item 20 also from the Remember subscale (remembering to follow the recommendations in case of bleeding) with an item-total correlation of .74 for the total sample. None of the items of the Plan subscale were correlated substantially to either their own subscale or to the total scale. It was also the case of item 1 (child report only) and item 3 of the Treat subscale, item 12 of the Dose subscale (parent report only), item 17 of the Remember subscale (parent report only), and item 22 of the Communicate subscale.

VERITAS-Pro and VERITAS-PRN Scores

Mean scores are presented in Tables 4 and 5. Total scores for the total sample ranged from 24 to 67 for the VERITAS-Pro and from 24 to 70 for the VERITAS-PRN. In general, the participants in our study had similar adherence scores to prophylaxis (*M* = 39.30 ± 12.10) as in the validation study (*M* = 41.2 ± 14.7, d = .14) (Duncan et al., 2010a). However, the parents of our sample, when they were the primary infusers, reported lower scores for prophylaxis (*M* = 31.00 ± 5.20, d = .50) than those in the validation study (35.3 ± 11.1), indicating higher adherence in our sample. Participants in our study also reported higher adherence for episodic treatment ($M = 36.88 \pm 10.56$) than in the validation study ($M = 46.5 \pm 11.1$, $d = 0.89$) (Duncan et al., 2010b). It is important to note that the participants of our study were not strictly on an episodic regimen as in the validation study because (they also received prophylaxis), and the validation study only included patients over the age of 16.

When they were primary infusers, children reported more skipping behaviors for prophylaxis (VERITAS-Pro, Skip subscale) than in parent estimates of children behaviors (Wilcoxon $Z = 2.032$, $p < .05$). They also tended to report lower adherence to episodic treatment (VERITAS-PRN, total score) than their parent estimates ($Z = 1.926$, $p < .10$). Also, whether the child ($Z = 1.826$, $p < .10$) or the parent ($Z = 2.132$, $p < .05$) was the primary infuser, children tended to report lower adherence for the Time subscale vs. parents' report for episodic treatment (VERITAS-PRN, Time subscale).

Moreover, parents rated higher adherence on prophylaxis when they were themselves in charge of performing the infusion than when children were autonomous with infusions (Mann-Whitney $U = 11.50$, $Z = -2.895$, $p < .01$). In children's report, this difference was not significant.

When comparing levels across domains of adherence within each instrument, the highest adherence was on the Dose subscale for prophylaxis (total sample, Friedman's ANOVA, $p < .01$; Wilcoxon, $p < .05$). For episodic treatment, the lowest adherence was on the Communicate subscale (total sample, Friedman's ANOVA, $p < .01$; Wilcoxon, $p < .05$).

Last year bleeding episodes and adherence

A significant correlation was found between adherence as reported by children and the number of bleeding episodes in the last year ($r = -.73$, $p < .01$ with VERITAS-Pro scores, $r = -.63$, $p < .05$ with VERITAS-PRN scores). Counterintuitively, children experiencing more bleeding episodes in the last year reported higher adherence ratings. We will discuss this result and the possible role of behavior change in the next section. Interestingly, there were no significant associations when parents' estimates were considered ($r = -.22$, $p = .36$ with VERITAS-Pro scores, $r = -.09$, $p = .72$ with VERITAS-PRN scores).

To obtain the French-language versions, contact the author of the original questionnaires, Nathalie A. Duncan (email: nduncan@ihtc.org).

Discussion

In hemophilia, the lack of a validated measure of adherence to treatment has been a barrier to adherence research in the French-speaking population. We translated into French the VERITAS-Pro and VERITAS-PRN using a standard procedure based on understandability by the pediatric population. We also reported internal consistency of both instruments in an independent pediatric sample. The questionnaires were adapted to French without major issues. Results of this preliminary study support the reliability of the French versions of the tools, especially when using total scores. However, the results suggest there may be difficulties with the internal consistency of the Dose subscale of the VERITAS-Pro and the Plan subscale of the VERITAS-PRN in our population.

During the translation process, we achieved a French version that was easily understood by both pediatric patients and their parents. Consistency was particularly low for the Dose subscale of the VERITAS-Pro. It is possible to think that all items might not have been interpreted the same way by parents or might not reflect the practice at our healthcare center. For example, taking a close look at specific items, we found that although all patients and parents reported always using the right dose, as many as 36% of our total sample answered that they increased or decreased the dose without calling the hemophilia center. In our treatment center and under certain circumstances, healthcare professionals may advise some patients to add an infusion or to slightly increase the dose prior to some specific physical activity. For those parents, positive answers may not represent a lack of adherence. Furthermore, none of the items of the Dose subscale were associated with either the total scale or the subscale score. It is possible that the low variability in scores impacted the size of the correlations as most participants judged their adherence to be high on Dose issues (with all of the participants answering being fully adherent to item 5).

The Plan subscale also had low internal consistency for both VERITAS-Pro (parent report) and VERITAS-PRN (parent and child report). It should be noted that the child report for this subscale might be biased because the management of supplies may well be under the parent’s responsibility without them necessarily being aware of their parent’s behaviors regarding those aspects. Furthermore, low variability in scores may have impacted the size of the correlations, as adherence was generally good regarding planning aspects.

Although the main objective of this study was to translate the instrument and present the preliminary psychometric characteristics of the translated version, we nonetheless present certain descriptive results relating to the estimation of adhesion in our sample. These data should be interpreted with caution given the sample size, as well as the fact that the validation of these versions is in its early stages.

The VERITAS-Pro and VERITAS-PRN provided a detailed measurement of adherence. It thus may help identify areas where adherence could be improved. Communicating with the treatment center in case of questions or concerns received the lowest estimate of adherence for episodic treatment and showed some of the lowest adherence scores for prophylaxis. Working on establishing a good relationship with patients and their parents in order to have them willingly call their hemophilia center might be a key to increase adherence in pediatric patients. Time spent with the healthcare providers and good relationships with the nurse and the hematologist are factors associated with increased adherence in hemophilia (De Moerloose, Urbancik, Van Den Berg, & Richards, 2008; Tran et al., 2017) On the other hand, the Dose subscale received the highest adherence score for prophylaxis, just as it was the case in the validation study of the VERITAS-Pro (Duncan et al., 2010a). For the communication subscale, the standard deviation was the largest of all subscales in both questionnaires, which could be explained by large individual differences on those aspects. However, we also noticed that item 24 could lend itself to different interpretations. The wording "I call the treatment center when I have questions …" might have led some participants to answer about the occurrence of their questions rather than their reliance on their health care team when they have questions. We noticed the same issue in the original version.

In a worldwide survey, infrequent bleeding was ranked as the most common reason for nonadherence (Geraghty et al., 2006b). In our study, we found that children's total scores positively correlated with past year bleeding episodes which could indicate that having serious bleeding episodes might act as a motivator for future adherence. Similar results were found in a Spanish validation of the VERITAS-Pro on 13-62 year-old patients in which those who have had bleeding episodes in the last month had higher adherence (Cuesta-Barriuso et al., 2017). According to our results, this increased adherence could persist in time, even after the full recovery of hemarthrosis. However, in our sample, parent report of adherence was not correlated with the number of bleeding episodes in the last year. It is possible that the way children answered the questionnaire was more influenced by their recent bleeding episodes or that parents were not always aware of their child's behavior regarding adherence, especially when children were more autonomous with their injection.

Pediatric patients who were autonomous with their treatment also had a lower adherence (as reported by parents) compared to children whose parents were in charge of infusing. These results are consistent with studies finding higher adherence to treatment at a younger age when parents are the primary infusers (Duncan, Shapiro, Ye, Epstein, & Luo, 2012; Geraghty et al., 2006a). Decreased adherence at adolescence is common in many chronic illnesses with explanations ranging from emphasis on the present, inability to see the long-term consequences, testing limits, and so on (Taddeo, Egedy, & Frappier, 2008a). In our study, there was also a pattern for children to report lower adherence than in parents' estimates, but the differences were not significant across all scales. This could mean that parents are not always aware of all instances of nonadherence from their older children, or that children underestimated their degree of adherence, basing their opinion on recent events of nonadherence. It was also reported that parents of children with hemophilia experienced significant levels of guilt as compared to parents of healthy children, e.g., guilt over genetic transmission(Browne, Mally, & Kane, 1960; Kim, Kang, Cho, Song, & Ji, 2008). It is possible that this guilt makes them less inclined to report events of nonadherence. This might have been particularly the case for the Time subscale of the VERITAS-PRN, which refers to infusing immediately when there are signs of bleeding instead of waiting for a more convenient moment. For this subscale, parents' estimates tended to reflect systematically higher adherence than in the children's reports, no matter who were the primary infusers. This is very consistent with the common observation in child psychology, that it is necessary to explore children's behaviors through the views of both, children and parents.

We should recognize the limitations of this research. Firstly, the external validity of our results is limited due to the prophylaxis treatments being available and covered by the healthcare system in Canada, which may not be the case in other French-language areas, such as Africa or the Caribbean. Secondly, the small sample size limited the scope of psychometric analyses and also prevented us from studying dimensionality using standard procedures like factorial analysis. Further studies should explore the consistency and validity of these adaptations of VERITAS-Pro and VERITAS-PRN in larger samples.

To conclude, we developed a French-language adaptation of questionnaires to assess adherence in hemophilia VERITAS-Pro and VERITAS-PRN. We confirmed feasibility and understandability in a pediatric population using patient report and parent report. Preliminary psychometric analyses support the use of the scales with good to excellent consistency for total scores in both questionnaires. However, concerns were raised about some subscales that may lack consistency. Perhaps individual items should be adapted to increase consistency. Future studies should address the reliability and validity of the scales in a French population.

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Declaration of interest

The authors declared no conflict of interest

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Table 1. Demographic sample characteristics

	Total	
	Child report *n* = 13	Parent report *n* = 20
Age in years, mean (SD)	13 (3)	12 (4)
Hemophilia type n (%)		
A	11 (85)	17 (85)
B	2 (15)	3 (15)
Country of birth n (%)		
Canada	12 (92)	17 (85)
Other	1 (8)	3 (15)

Note: Mean age of children who were primary infusers was 14 ± 3 yrs for prophylaxis and 16 ± 3 yrs for episodic treatment.
Mean age of children whose parents were primary infusers was 10 ± 2 yrs for prophylaxis and 10 ± 3 yrs for episodic treatment.

Table 2. VERITAS-Pro internal consistency: inter-item correlations and Cronbach's alphas

	Inter-item correlation			Cronbach's α		
	All *n* = 33	Child report *n* = 13	Parent report *n* = 20	All *n* = 33	Child report *n* = 13	Parent report *n* = 20
Total scale	.29	.37	.22	.89	.91	.83
Time	.46	.50	.41	.75	.77	.71
Dose	.05	.09	.03	.13	.28	.09
Plan	.33	.48	.13	.61	.76	.37
Remember	.52	.59	.48	.80	.84	.73
Skip	.64	.67	.60	.87	.89	.78
Communicate	.30	.27	.32	.65	.61	.69

Note: Low values are underlined (inter-item correlation <.15; Cronbach's alpha <.30)

Table 3. VERITAS-PRN internal consistency: inter-item correlations and Cronbach's α

	Inter-item correlation			Cronbach's α		
	All *n* = 33	Child report *n* = 13	Parent report *n* = 20	All *n* = 33	Child report *n* = 13	Parent report *n* = 20
Total scale	.22	.22	.18	.85	.87	.73
Treat	.32	.27	.61	.68	.62	.73
Time	.40	.33	.34	.69	.64	.60
Dose	.42	.53	.30	.66	.78	.41
Plan	.02	.10	-.05	.01	.23	-.28
Remember	.51	.57	.33	.76	.82	.53
Communicate	.33	.39	.15	.64	.70	.37

Note: Low values are underlined (inter-item correlation <.15; Cronbach's alpha <.30)

Table 4. Mean and standard deviation for VERITAS-Pro scores

Mean (SD)	Total			Children as primary infusers		Parents as primary infusers	
	All *n* = 33	Child report *n* = 13	Parent report *n* = 20	Child report *n* = 9	Parent report *n* = 9	Child report *n* = 4	Parent report *n* = 11
Total scale	39.30 (12.10)	44.62 (14.07)	35.85 (9.48)	47.56 (15.59)	41.78 (10.38)	38.00 (7.75)	31.00 (5.20)
Time	6.27 (2.39)	7.00 (2.45)	5.80 (2.29)	7.67 (2.60)	7.22 (2.68)	5.50 (1.29)	4.64 (0.92)
Dose	4.97 (1.38)	4.85 (0.99)	5.05 (1.61)	5.00 (1.00)	4.78 (0.83)	4.50 (1.00)	5.27 (2.05)
Plan	6.61 (2.62)	7.38 (3.15)	6.10 (2.15)	7.89 (3.62)	6.89 (2.71)	6.25 (1.50)	5.45 (1.37)
Remember	7.21 (3.09)	8.62 (3.31)	6.30 (2.64)	9.11 (3.72)	8.11 (2.37)	7.50 (2.08)	4.82 (1.83)
Skip	6.06 (3.05)	7.54 (3.48)	5.10 (2.36)	7.78 (4.02)	5.67 (3.24)	7.00 (2.16)	4.64 (1.29)
Communicate	8.18 (3.55)	9.23 (4.09)	7.50 (3.07)	10.11 (4.40)	9.11 (2.89)	7.25 (2.75)	6.18 (2.64)

Table 5. Mean and standard deviation for VERITAS-PRN scores

Mean (SD)	Total			Children as primary infusers		Parents as primary infusers	
	All *n* = 33	Child report *n* = 13	Parent report *n* = 20	Child report *n* = 4	Parent report *n* = 4	Child report *n* = 9	Parent report *n* = 16
Total scale	36.88 (10.56)	42.69 (12.53)	33.10 (7.10)	48.25 (14.67)	34.00 (5.47)	40.22 (11.52)	32.88 (7.59)
Treat	4.94 (1.39)	5.31 (1.49)	4.70 (1.30)	5.25 (0.50)	4.75 (0.96)	5.33 (1.80)	4.69 (1.40)
Time	6.52 (2.67)	8.15 (2.82)	5.45 (1.99)	9.00 (2.31)	5.25 (1.26)	7.78 (3.07)	5.50 (2.16)
Dose	5.91 (2.61)	6.23 (3.44)	5.70 (1.98)	8.50 (5.80)	6.25 (1.71)	5.22 (1.20)	5.56 (2.06)
Plan	5.33 (1.83)	5.77 (2.09)	5.05 (1.64)	6.00 (1.83)	4.00 (0.00)	5.67 (2.29)	5.31 (1.74)
Remember	6.00 (2.69)	7.15 (3.48)	5.25 (1.74)	8.25 (4.35)	5.50 (2.38)	6.67 (3.20)	5.19 (1.64)
Communicate	8.18 (3.51)	10.08 (4.01)	6.95 (2.56)	11.25 (2.99)	8.25 (2.87)	9.56 (4.25)	6.63 (2.47)

Supplementary File 1. VERITAS-Pro Mean (SD), item-total and item-subscale correlations for the total sample, child report and parent report

		All *n* = 33			Child report *n* = 13			Parent report *n* = 20		
Subscale	Items	Mean (SD)	Item-total	Item-subscale	Mean (SD)	Item-total	Item-subscale	Mean (SD)	Item-total	Item-subscale
Time	Item 1	1.39 (.61)	.65	.71	1.54 (.52)	.75	.68	1.30 (.66)	.62	.73
	Item 2	1.27 (.63)	.63	.41	1.38 (.51)	.62	.45	1.20 (.70)	.69	.38
	Item 3	1.91 (1.01)	.57	.53	2.23 (1.01)	.75	.76	1.70 (.98)	.34	.36
	Item 4	1.70 (.85)	.68	.62	1.85 (.99)	.73	.60	1.60 (.75)	.63	.63
Dose	Item 5	1.00 (.00)	-	-	1.00 (.00)	-	-	1.00 (.00)	-	-
	Item 6	1.21 (.74)	-.07	.15	1.15 (.38)	.21	-.09	1.25 (.91)	-.15	.20
	Item 7	1.55 (.79)	.23	.17	1.46 (.66)	.45	.42	1.60 (.88)	.19	.13
	Item 8	1.21 (.74)	.06	-.08	1.23 (.44)	.39	.29	1.20 (.89)	-.09	-.14
Plan	Item 9	1.45 (.67)	.53	.41	1.77 (.83)	.61	.57	1.25 (.44)	.20	.10
	Item 10	1.64 (.96)	.51	.52	1.85 (.99)	.67	.81	1.50 (.95)	.31	.23
	Item 11	1.48 (.62)	.66	.37	1.69 (.63)	.75	.41	1.35 (.59)	.52	.27
	Item 12	2.03 (1.40)	.46	.43	2.08 (1.50)	.58	.60	2.00 (1.38)	.39	.30
Remember	Item 13	1.97 (.88)	.73	.71	2.46 (.78)	.63	.58	1.65 (.81)	.78	.78
	Item 14	1.70 (.92)	.78	.72	2.08 (.95)	.80	.78	1.45 (.83)	.72	.60
	Item 15	1.82 (1.07)	.52	.40	1.85 (.99)	.83	.72	1.80 (1.15)	.38	.26
	Item 16	1.73 (1.01)	.65	.69	2.23 (1.24)	.63	.69	1.40 (.68)	.56	.64
Skip	Item 17	1.76 (1.09)	.54	.62	2.08 (1.12)	.69	.91	1.55 (1.05)	.32	.30
	Item 18	1.45 (.83)	.72	.86	1.85 (.90)	.67	.89	1.20 (.70)	.70	.78
	Item 19	1.42 (.83)	.59	.72	1.77 (.93)	.34	.61	1.20 (.70)	.80	.78
	Item 20	1.42 (.83)	.67	.72	1.85 (1.07)	.53	.65	1.15 (.49)	.80	.78
Communicate	Item 21	2.39 (1.52)	.59	.74	2.92 (1.75)	.73	.65	2.05 (1.28)	.31	.82
	Item 22	2.00 (1.23)	.57	.59	2.31 (1.49)	.68	.51	1.80 (1.01)	.35	.65
	Item 23	2.21 (1.19)	.44	.34	2.15 (1.28)	.53	.36	2.25 (1.16)	.46	.39
	Item 24	1.58 (1.09)	-.04	.14	1.85 (1.46)	-.19	.11	1.40 (.75)	.03	.10

Note: item 5 had no variation, all participants gave the highest rating of adherence

Supplementary File 2. VERITAS-PRN Mean (SD), item-total and item-subscale correlations for the total sample, child report and parent report

		All *n* = 33			Child report *n* = 13			Parent report *n* = 20		
Subscale	Items	Mean (SD)	Item-total	Item-subscale	Mean (SD)	Item-total	Item-subscale	Mean (SD)	Item-total	Item-subscale
Treat	Item 1	1.24 (.44)	.28	.40	1.23 (.44)	.19	.26	1.25 (.44)	.51	.56
	Item 2	1.30 (.59)	.59	.67	1.54 (.66)	.41	.58	1.15 (.49)	.70	.77
	Item 3	1.06 (.24)	.29	.22	1.15 (.38)	.21	.28	1.00 (.00)	-	-
	Item 4	1.33 (.60)	.65	.64	1.38 (.65)	.73	.54	1.30 (.57)	.69	.74
Time	Item 5	1.55 (.83)	.67	.60	1.85 (.99)	.74	.58	1.35 (.67)	.45	.52
	Item 6	1.97 (1.24)	.36	.59	2.69 (1.44)	.31	.47	1.50 (.83)	.01	.51
	Item 7	1.21 (.42)	.48	.46	1.31 (.48)	.36	.37	1.15 (.37)	.59	.49
	Item 8	1.79 (1.02)	.55	.43	2.31 (.95)	.62	.41	1.45 (.95)	.32	.22
Dose	Item 9	1.15 (.36)	.48	.51	1.23 (.44)	.35	.47	1.10 (.31)	.65	.55
	Item 10	1.33 (.82)	.58	.71	1.38 (1.12)	.59	.80	1.30 (.57)	.74	.55
	Item 11	1.36 (.99)	.46	.44	1.38 (1.12)	.68	.80	1.35 (.93)	.32	.05
	Item 12	2.06 (1.30)	.41	.37	2.23 (1.48)	.57	.51	1.95 (1.19)	.20	.19
Plan	Item 13	1.30 (.81)	.11	.02	1.38 (.65)	.12	.21	1.25 (.91)	.07	-.12
	Item 14	1.21 (.74)	.11	.23	1.46 (1.13)	-.06	.38	1.05 (.22)	.20	.00
	Item 15	1.24 (.75)	-.05	-.07	1.23 (.44)	-.17	.13	1.25 (.91)	-.00	-.15
	Item 16	1.58 (1.25)	.29	-.09	1.69 (1.32)	.53	-.10	1.50 (1.24)	.04	-.12
Remember	Item 17	1.27 (.63)	.56	.58	1.54 (.88)	.58	.66	1.10 (.31)	.24	.18
	Item 18	1.42 (.83)	.61	.48	1.46 (1.13)	.72	.53	1.40 (.60)	.58	.46
	Item 19	1.88 (1.24)	.58	.51	2.38 (1.33)	.63	.60	1.55 (1.10)	.38	.28
	Item 20	1.42 (.71)	.74	.85	1.77 (.93)	.68	.89	1.20 (.41)	.73	.70
Communicate	Item 21	2.12 (1.17)	.67	.69	2.77 (1.24)	.66	.80	1.70 (.92)	.51	.40
	Item 22	2.27 (1.44)	.50	.29	2.92 (1.50)	.64	.16	1.85 (1.27)	.11	.22
	Item 23	2.24 (1.30)	.26	.36	2.46 (1.33)	.31	.62	2.10 (1.29)	.15	.08
	Item 24	1.55 (1.12)	.25	.42	1.92 (1.44)	.14	.49	1.30 (.80)	.16	.14

Note: item 3 for parent report had no variation, all participants gave the highest rating of adherence

Introduction à l’article 2

À cette étape de la thèse, nous voulions d’abord déterminer si les enjeux d’adhésion étaient plus importants en ce qui a trait au traitement par injections ou en ce qui concerne l’activité physique, afin d’orienter le programme de thèse vers ce qui semble le plus problématique et le plus pertinent pour l’intervention en milieu hospitalier. Ainsi, l’article 2 s’insérait dans l’objectif premier de la thèse, soit de décrire le phénomène de l’adhésion chez les jeunes atteints d’hémophilie.

Ce deuxième article visait également à répondre au deuxième objectif général de la thèse, soit d’étudier une première catégorie de facteurs possiblement liés à la motivation des jeunes à suivre les recommandations émises par leur équipe soignante, tirés ici de la théorie du comportement planifié. L’article 2 visait donc à répondre aux questions suivantes : les jeunes atteints d’hémophilie sont-ils motivés par leur traitement, ont-ils l’intention de suivre les recommandations émises par leur équipe de soin ? Voient-ils les recommandations comme étant somme toute positives ou négatives ? Pensent-ils que ces mesures peuvent les aider à prévenir les saignements articulaires ? Se sentent-ils capables de suivre toutes les mesures ? Sont-ils influencés par leurs pairs et par leurs parents en ce qui a trait à leur adhésion?

Dans l’article 2, nous avons fait le choix de nous concentrer davantage sur l’étude de la motivation des jeunes (leur intention) plutôt que sur leur comportement d’adhésion. La raison qui nous a amené à prendre cette décision est que la plupart des jeunes ne sont pas totalement libres de leurs actions. En effet, l’encadrement des parents et du personnel de l’école fait en sorte que le comportement des jeunes n’est pas nécessairement représentatif de la conduite qu’ils adopteraient s’ils étaient plus autonomes (ou de comment ils agissent lorsqu’ils sont moins surveillés). Notre intérêt se portait donc davantage sur leur motivation personnelle face aux comportements de prévention des saignements. Il est crucial que les jeunes développent et maintiennent cette motivation au cours de la transition vers l’âge adulte.

Étudier l’intention de suivre les recommandations du plan de traitement chez jeunes atteints d’hémophilie est particulièrement important étant donné la baisse de leur adhésion durant la transition vers l’autosoin. Si les jeunes n’ont pas l’intention de suivre les recommandations, on peut penser que faibles sont les chances qu’ils les suivront lorsqu’ils seront moins supervisés par leurs parents. Plusieurs études empiriques ont en effet démontré un lien étroit entre l’intention et le comportement dans le domaine de la santé (Godin & Kok, 1996; Sheeran, 2002). Une méta-analyse de méta-analyses a révélé que l'intention d'adopter un comportement expliquait en moyenne 28% de la variance du comportement (Sheeran, 2002). Bien qu'insuffisante à elle seule pour prédire parfaitement l'adhésion, la TCP est l'une des théories les plus performantes et les plus utilisées pour identifier les facteurs qui maintiennent ou entravent changements comportementaux en santé (Ajzen, 1991). La TCP explique que les individus détermineraient à l'avance leurs intentions concernant la plupart de leurs comportements volontaires et que l'intention serait le meilleur prédicteur de leur comportement futur (Sarafino, 2011). Ainsi, l'intention peut être conceptualisée comme une mesure du degré de motivation personnelle des jeunes pour leur traitement.

Trois facteurs principaux que nous décrirons plus loin seraient à l'origine des intentions : l'attitude, la norme subjective et la perception de contrôle comportemental (voir figure du modèle complet à l'Annexe A). Selon une revue de la littérature sur l'efficacité de cette théorie pour prédire les comportements en santé, l'attitude, la norme subjective et la perception de contrôle comportemental expliquaient en moyenne 41% de la variance de l'intention et 34% de la variance du comportement (Godin & Kok, 1996). Une étude sur la prédiction de l'activité physique chez l'adolescent a démontré que les facteurs de la TCP expliquaient 56% de la variance de l'intention (Foley et al., 2008).

La théorie explique que l'attitude, la norme subjective et la perception de contrôle comportemental seraient des construits intrapsychiques qui se formeraient, eux, à partir d'un ensemble de croyances qui sont, elles, déterminées par une multitude de facteurs individuels et sociaux, comme la personnalité et l'éducation. L'étude de ces facteurs permettrait le développement d'interventions efficaces pour favoriser l'autosoin et l'activité physique (Godin & Kok, 1996; Lin et al., 2019; Parrott, Tennant, Olejnik, & Poudevigne, 2008; Sanaeinasab et al., 2019).

L'*attitude* est un des prédicteurs proximaux de l'intention. L'attitude peut être définie comme une tendance de l'individu à répondre favorablement ou non à un comportement spécifique (Fishbein & Ajzen, 2011). Il s'agirait d'une évaluation consciente et inconsciente basée sur les conséquences auxquelles une personne s'attend suite à un comportement donné (les croyances concernant les coûts et bénéfices d'un comportement), ainsi que l'importance ou la valeur donnée à ces conséquences (Tirado Gonzales, Neipp Lopez, Quiles Marcos, & Rodriguez-Marin, 2012). Par exemple, l'attitude envers l'activité physique sécuritaire représente d'une part le résultat attendu par la personne si elle pratique ce type d'activités (p.ex. une diminution du nombre de saignements articulaires), et d'autre part, la valeur de ce résultat à savoir s'il représente un objectif important de la personne. Selon la théorie, il serait possible d'obtenir une mesure dite directe de l'attitude, donc de la valence positive ou négative de celle-ci en utilisant plusieurs échelles sémantiques de type utile/nuisible, agréable/désagréable, positif/négatif (Fishbein & Ajzen, 2011). Il existerait aussi des mesures indirectes de l'attitude plus axées sur les croyances à propos des risques et bénéfices d'un comportement donné (Fishbein & Ajzen, 2011).

Identifier si l'attitude des jeunes atteints d'hémophilie envers leur plan de traitement influence leur intention de suivre les recommandations permettrait notamment de confirmer ou d'infirmer la nécessité d'informer davantage les jeunes à propos des risques et des bénéfices de chacun des comportements recommandés et déconseillés. Dans une étude incluant des participants dans 147 centres de traitements à travers le monde, la raison de non-adhésion la plus citée par les répondants (75% de ceux-ci) était l'incapacité à comprendre les bénéfices du traitement (Geraghty et al., 2006b). Dans une revue de la littérature dans le domaine de la santé, l'attitude est ressortie comme prédicteur significatif dans 82% des études avec une corrélation moyenne avec l'intention de $r = 0.46$ (Godin & Kok, 1996). Par contre, à ce jour, nous ne savons pas dans quelle mesure l'attitude affecte l'intention et l'adhésion réelle des jeunes patients atteints d'hémophilie. Les jeunes ayant tendance à mettre l'accent sur le présent, il est possible qu'ils ne tiennent pas compte des risques et des bénéfices lorsque vient le temps de choisir une activité physique par exemple, mais qu'ils soient plutôt influencés par l'opinion de leurs amis.

La *norme subjective* représente la perception qu'un individu a à savoir si les personnes qui sont importantes pour lui pensent qu'il devrait ou non adopter un comportement (Sarafino, 2011). En d'autres mots, elle représente le degré avec lequel une personne sent que les autres (amis, parents, autres membres de la société) l'incitent à s'engager ou non dans un comportement donné. La norme subjective comprend également la motivation de la personne à se conformer ou non à l'opinion des autres.

Dans une étude sur les enfants atteints de maladies chroniques, le support de la part des parents et des amis a été démontré comme un prédicteur significatif de l'adhésion au traitement chez les jeunes. En effet, ceux-ci étaient 2.69 fois plus susceptibles d'être adhérents s'ils recevaient le support de leurs parents et 2.11 fois plus susceptibles si leurs amis les supportaient, que s'ils ne recevaient pas cet appui (Kyngas & Rissanen, 2001). Dans une autre revue systématique sur le diabète de l'adolescent, une maladie dont l'autosoin implique aussi de recevoir des injections, l'influence parentale est ressortie comme étant le facteur le plus important de non-adhésion au traitement (Cox & Hunt, 2015). Pour ce qui est de l'activité physique, le support des autres et des parents est un facteur qui revient de manière constante comme prédicteur du comportement (Foley et al., 2008; Sallis et al., 2000). Chez les adolescents atteints de diabète par exemple, être encouragé par les autres à s'engager dans des comportements contraires à ce qui est recommandé a été mentionné comme étant un facteur important de non-adhésion (Palladino & Helgeson, 2013). Bien que peu d'études portent sur l'influence des pairs et de l'environnement familial en hémophilie, nous avons identifié une étude montrant que les familles les moins adhérentes trouvaient difficile d'administrer la prophylaxie dans leur contexte familial ou social (Hacker et al., 2001). Un des avantages importants de la TCP dans l'étude des comportements de santé est qu'elle prend en compte l'influence sociale sur le comportement.

Le *contrôle perçu* concerne la confiance qu'un individu possède concernant ses capacités, ses connaissances et ses habiletés pour réaliser un comportement donné (Fishbein & Ajzen, 2011). Le concept de contrôle perçu tel que décrit dans la TCP s'apparenterait au concept d'autoefficacité de Bandura (Bandura, 1977; Fishbein & Ajzen, 2011). Certains auteurs tenteraient de les différencier, mais pour d'autres, ces deux concepts seraient identiques (Fishbein & Ajzen, 2011). Le contrôle comportemental perçu comporte deux sous-éléments, soit l'impression de capacité ou de maîtrise de l'individu face au comportement, ainsi que son autonomie, c'est-à-dire le degré avec lequel il est libre d'exercer ce comportement (Fishbein & Ajzen, 2011). Le contrôle perçu est très important dans la prédiction de l'intention comportementale (Tirado Gonzales et al., 2012). Dans le domaine de la santé, il a été la cible de plusieurs interventions efficaces (Schwarzer & Fuchs, 1996). Dans une revue de la littérature dans le domaine de la santé, le contrôle perçu est ressorti comme prédicteur significatif de l'intention d'accomplir les comportements de santé dans 85,5% des études avec une corrélation moyenne de $r = 0.46$ (Godin & Kok, 1996).

En plus des prédicteurs proposés par la TCP directement, plusieurs autres facteurs ont été proposés dans d'autres études afin d'augmenter la valeur prédictive du modèle, mais ces facteurs différeraient en fonction du comportement étudié et ne pourraient donc pas être intégrés à la théorie générale (Fishbein & Ajzen, 2011). Un des facteurs qui a souvent présenté de fortes corrélations avec l'intention et le comportement futur est le comportement passé de l'individu

(Fishbein & Ajzen, 2011; Ouellette & Wood, 1998). Selon les auteurs de la TCP, l'inclusion du comportement passé serait en effet susceptible d'expliquer davantage de variance, mais le lien entre le comportement passé et futur serait indirect et en lien avec des facteurs intrapsychiques qui restent à définir avant de pouvoir être inclus dans la théorie (Fishbein & Ajzen, 2011). Nous avons tout de même décidé d'inclure le comportement passé dans notre étude en plus des facteurs mentionnés plus haut.

Ce deuxième article visait à identifier les facteurs sociocognitifs issus de la théorie du comportement planifié qui sont liés à l'intention de se comporter selon les recommandations médicales pour les domaines suivants : 1) le traitement par injection prophylactique 2) le traitement par injection sur demande, 3) la pratique d'activité physique recommandée et 4) la pratique d'activité physique déconseillée. Pour ce faire, nous avons mesuré les associations entre l'intention de suivre les recommandations pour chacun de ces quatre types de comportements et les facteurs suivants : l'attitude (l'attitude générale et l'attitude spécifique en lien avec la prévention des saignements), la norme subjective (reliée aux parents et aux amis), le contrôle perçu, et le comportement passé (l'adhésion au traitement dans le passé et la pratique d'activité physique dans le passé).

De façon cohérente avec la littérature, nous nous attendions à ce qu'une plus grande intention de pratiquer de l'activité physique recommandée soit associée à une attitude générale plus positive envers l'activité physique recommandée, à une perception d'un environnement qui encourage davantage ce comportement et à une plus grande impression de contrôle. De la même façon, nous nous attendions à ce qu'une intention plus forte de pratiquer de l'activité physique déconseillée soit associée à une perception d'un environnement qui appuie davantage ce comportement, mais également à une attitude selon laquelle le jeune ne voit pas ou n'accorde pas d'importance au risque de saignements associé à ces activités. De même, nous nous attendions à ce qu'une intention accrue de respecter les recommandations au sujet du traitement prophylactique et du traitement sur demande soit expliquée par une attitude plus positive par rapport à l'efficacité de ces mesures à prévenir les saignements et à un meilleur contrôle comportemental.

Article 2

Understanding adherence to treatment and physical activity in children with hemophilia: The role of psychosocial factors

Sarah Bérubé[1,2], Audrey Cloutier-Bergeron[3], Claudine Amesse[2], and Serge Sultan[1,2*]

[1] Université de Montréal, Québec, Canada
[2] CHU Sainte-Justine, Montréal, Québec, Canada
[3] Université Laval, Québec, Canada

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Abstract

The objective of this study was to identify psychosocial factors to explain intentions of children and adolescents with hemophilia to adhere to recommendations for self-care. Twenty-four patients with hemophilia A and B, aged 6-18 years old, and their parents, completed a survey. Measures assessed factors from the theory of planned behavior, physical activity, and medical treatment adherence. The results indicate that past behaviors, attitudes, and subjective norms explained a large proportion of the intention to engage in future non-recommended physical activity. This study supports the need to investigate motivational factors underlying behaviors in larger-scale studies and identifies targets for future interventions.

Keywords: Adherence; Hemophilia; Intention; Physical Activity; Theory of Planned Behavior

Introduction

Hemophilia is a rare genetic disorder that primarily affects men and that is caused by a clotting factor deficiency, leading to bleeding episodes in muscles and joints. Bleeds not immediately treated may eventually lead to impairment such as arthritis or disability (Nazzaro et al., 2006). Hemophilia can be medically managed with a lifelong prophylactic or on-demand treatment, which involves the infusion of the deficient clotting factor to help prevent bleeding and arthropathy. Regular exercise is recommended to patients in order to promote strong muscles, develop balance and improve fitness, which in return can contribute to healthier joints and diminish the occurrence of bleeds. However, physical activity likely to cause trauma, such as high contact and collision sports, should be avoided (Srivastava et al., 2013a).

Like in other chronic illnesses, difficulties in following recommendations for self-care have been reported in children and adolescents with hemophilia for both physical activity and treatment adherence. While suboptimal adherence is not uncommon in parents of young people living with hemophilia, adherence tends to be higher when parents take responsibility of their children's infusion (Schrijvers et al., 2016). When young people with hemophilia gain in autonomy and start regularly self-infusing, adherence to treatment reduces remarkably. In a recent survey, 90% of children aged 0-12 years old had a "high" or "very high" adherence to prophylaxis, compared with 54% for those aged 13-18 years old (Geraghty et al., 2006b). In addition, despite recommendations, a survey revealed that 60% of young people with hemophilia manage their condition by limiting any type of physical activity (Nazzaro et al., 2006). Other studies suggest that a high proportion of children and youth with hemophilia engage in high-risk sports just like their friends (Mulder et al., 2004; C. Ross et al., 2009).

Studies on adherence of people with hemophilia have identified over 20 reasons, mostly provided by healthcare professionals or patients, to explain nonadherence (ie, "lack of time", "forgetfulness", "fluctuation or disappearance of symptoms", etc.), although these reasons were not systematically correlated with the actual level of adherence (De Moerloose et al., 2008; Geraghty et al., 2006b; Hacker et al., 2001; Nazzaro et al., 2006; Schrijvers et al., 2013). However, no theoretical model has been used until now to understand motivation for treatment, thus precluding the development of intervention strategies.

The theory of planned behavior is one of the most frequently used theories to understand motivation for treatment, and posits that intention to adopt a behavior is a central predictor of future behavior (Ajzen, 1991). Studying intention allows us to study a child's level of motivation to their treatment plan and its determinants. In the pediatric context, the study of behavior is complicated by the fact that young people are under the supervision of their parents. Thus, their behavior does not necessarily represent how they would behave if they were more independent or when they are not in the presence of their parents. To counter this problem, it is possible to assess a child's intention to follow future recommendations. The theory of planned behavior posits three core factors leading to the formation of intention: attitudes, subjective norms and perceived behavioral control. Finally, past behavior can also influence intentions and behaviors (Ouellette & Wood, 1998).

According to this theory*, attitudes* are based on the patient's beliefs about the consequences of performing the behavior (i.e., costs and benefits) and can be assessed broadly or by its specific components (e.g., the anticipated benefits of the recommendations in preventing joint bleeds) (Fishbein & Ajzen, 2011). *Subjective norm* represents the social influence on behavior and is based on the patient's beliefs about others' opinions of the behavior, and the patient's motivation to comply with these opinions. Finally, *perceived behavioral control* refers to the confidence about one's abilities, knowledge and skills to perform the behavior. The theory of planned behavior has been widely used to predict many health-related behaviors in people of all ages, and successful interventions have been developed using this model (Godin & Kok, 1996). To date, self-care in children and adolescents with hemophilia has not been studied through this framework. The identification of predictive factors of self-care could help identify important motivational targets for future interventions.

Objective

The objective of this study was to identify individual psychosocial factors to explain intentions of young people with hemophilia to adhere to the following recommendations: 1) infuse prophylactic treatment, 2) infuse on-demand treatment, 3) practice recommended physical activity, and 4) avoid practice of physical activity that was discouraged by their health care providers. To do so, we assessed associations between intentions to follow recommendations and potential psychosocial factors from the theory of planned behavior: attitudes towards recommendations, subjective norm (related to parents and friends), perceived behavioral control, and past behavior (past adherence to medication, recommended and discouraged physical activity).

Methods

Participants

Patients aged 6-18 years old with severe hemophilia A or B (VIII / IX < 1%), treated at our institution and accompanied by one of their parents, were approached for this study. We chose to recruit people with a more severe form of hemophilia because of the intensity of symptoms these patients experience and the use of prophylaxis as a treatment. All eligible patients spoke French or English. Exclusion criteria included: diagnosis < 1 year and presence of a severe psychiatric disorder. We excluded young patients who were diagnosed with a psychiatric disorder or had received their diagnosis less than a year before the study and were still adapting to the illness, in order to avoid outliers and collect answers from a relatively homogenous sample representative of the majority of young people with hemophilia. All thirty-one patients who were on the list of the local hemostasis centre and who met the criteria for inclusion and exclusion were approached for this study. Twenty-six patients (84%) and their parents accepted to participate (Table 1). Reasons for refusing to participate included: not having an appointment scheduled at the clinic ($n = 3$), and lack of time at the moment of the assessment ($n = 2$). Two participants were excluded from our study: one patient had an intellectual disability, and the other suffered from attention-deficit disorder, making a final sample size of 24. The mean age of participants was 11.8 ± 3.3 years old. All participants were on prophylaxis and received supplementary injections in case of bleeding (on-demand treatment).

The survey was a one-time assessment administered to participants during their biannual appointment at the hemophilia clinic. Questions were read to patients and a research assistant (SB) collected responses while one of their parents answered a questionnaire in a different room. This questionnaire included sociodemographic questions as well as questions on past behavior for younger children (see *Measures*). This project was approved by the Sainte-Justine UCH's Research Ethic Board.

Measures

To comply with theory of planned behavior norms and guidelines, we developed items and questions as recommended by Azjen and Fishbein (Ajzen, 2011; Fishbein & Ajzen, 2011; Fishbein, 2001).

Intention. Intention to adopt health behaviors was measured using one item for each behavior. The participant responded on a 7-level scale: *In the future, I intend to [the behavior] as recommended, 1-strongly disagree to 7-strongly agree*. For interpretative purposes, we reversed items on the intention to adopt discouraged physical activity, higher scores translating as the intention to avoid discouraged physical activity.

Attitudes. We measured general attitude towards health behaviors using a generic item: *I see* [the behavior] *as*: *1-negative to 7-positive*. We also evaluated the component of attitude related to the perceived benefits of the behavior in bleeding prevention by multiplying a measure of perceived benefit ([the behavior] *will prevent bleeding in my joints*, *1-strongly disagree to 7-strongly agree*) by a measure of perceived importance (*Preventing bleeding in my joints is*, *1-not important to 7-important*).

Subjective norms. Two forms of perceived social norms were measured: (a) perceived pressure from parents, and (b) from peers. Each social norm was measured using two items. The first assessed parents' or peers' perception of the behavior: *My friends appreciate that I [the behavior] as recommended, 1-strongly disagree to 7-strongly agree*; while the second assessed the degree of influence of the parents/peers: *Regarding my [behavior], I want to listen to my friends or do what they want me to do*, *1-strongly disagree to 7-strongly agree*.

Perceived behavioral control. Items of perceived behavioral control were also developed. They were phrased as*: How confident are you or do you feel able to always [the behavior] as recommended? 1-not at all confident to 7-very confident*.

Adherence to treatment was measured using the Validated Hemophilia Regimen Treatment Adherence Scale (VERITAS-Pro and VERITAS-PRN for adherence to prophylactic and on-demand treatment regimens respectively) (Bérubé, 2015; Duncan et al., 2010a; Duncan et al., 2010b). The questionnaires include 24 items and topics related to timing, dosing, planning, remembering, communication, and treating/skipping. Recent research indicated good psychometric properties - high reliability - with α values of .85 and .92 for total scale and a test-retest r > .77. The children's scores on the VERITAS questionnaires were used when the participants were autonomous in their treatment management. Otherwise, parent measures were used. In our sample, internal consistency for total scale remained high for VERITAS-Pro and VERITAS- PRN in both children-report (α values >.87) and parents-report (α values >.73).

To assess physical activity, we asked participants and parents to estimate physical activity in a typical week for both recommended and discouraged activities. As activities may vary greatly according to the season in Canada, we inquired for both winter and summer activities and averaged both sets of results. We used the same wording as in the Godin Leisure-Time questionnaire (Godin & Shephard, 1985): *How many days per week do you engage in recommended physical activity for at least 15 minutes?* Because estimation of physical activity is not reliable for children under 10 years old (Kohl, Fulton, & Caspersen, 2000), parent estimations were used with these participants.

Analysis

Associations between intention and psychosocial predictors were explored using partial correlation coefficients after controlling for age and past year bleeding episodes. Considering that our sample included a wide range of age, we used partial correlation to reduce systematic effect of age on the way participant could have understood and answered the questions. We also wanted to minimize the impact on the results that recent bleeding episodes can have on intentions to follow recommendations, as well as on attitudes, subjective norm, and perceived control, in the short-term. Nonparametric tests were conducted to compare levels of associations across behaviors (Wilcoxon signed-rank tests). Effect sizes of the differences (*r)* were also calculated and interpreted according to Cohen's convention of small ($r = 0.1$), medium ($r = 0.3$), and large ($r = 0.5$) effect sizes (Cohen, 1992). Finally, all variables (attitudes, subjective norm, and perceived control)were entered in a regression analysis for each of the four types of behavior and the model R-squared value was used to estimate the total variance of intentions explained by the studied psychosocial factors. An alpha level of .05 was used to detect significant associations. All analyses were performed with the *Statistical Package for the Social Sciences* SPSS v19.

Results

First, we observed that intention to follow recommendations was high for recommended physical activity, prophylaxis, and on-demand treatment (> 6/7 on a strongly disagree-strongly agree scale). However, intention to avoid practicing non-recommended physical activity was significantly lower than for other behaviors (Wilcoxon $Z = 2.31 - 2.96$, effect size $r = 0.33 - 0.47$, $p < .05$) (Table 2).

Practice of physical activity was higher for recommended activities (4.0 ± 1.9 days) as compared to that of discouraged activities (1.5 ± 1.9 days) ($Z = 3.429$, $r = 0.49$, $p < .05$). Importantly, practice of discouraged physical activity also significantly differed from 0 ($p < .05$). Adherence scores indicated relatively good adherence to prophylaxis and on-demand treatment from our participants in comparison to scores published in previous studies.

Attitudes were globally positive ($Z = 2.65 - 4.25$, $r = 0.38 – 0.61$, $p < .05$). However, attitude for avoiding risky physical activity was significantly lower than for other behaviors ($Z = 1.80 - 2.40$, $r = 0.28 – 0.35$, $p < .05$, except for on-demand treatment, $p = .07$) as was also the case for perceived behavioral control ($Z = 2.75 - 3.26$, $r = 0.39 – 0.52$, $p < .01$). Patients felt that their parent encouraged them less to follow the recommendations for physical activity than for other behaviors ($Z = 2.11 – 2.33$, $r = 0.32 – 0.37$, $p < .05$). Median perceived behavioral control for discouraged physical activity was neutral, indicating that participants as a group were not confident regarding their ability to avoid discouraged physical activity in the future. Concerning recommended physical activity, a neutral median score was found for the item pertaining to the utility of recommended physical activity in preventing bleeding episodes, meaning that participants did not necessarily see the usefulness of recommended physical activity as a preventative measure. To identify factors of behavioral intention, we focused on statistically significant partial correlations (Table 3). After controlling for age and bleeding episodes, higher intention regarding recommended physical activity tended to be associated with larger perceived encouragement from peers. Intention to avoid discouraged physical activity was associated with a lower practice of risky physical activity in the past, a positive general attitude toward avoidance, a positive attitude regarding the efficacy of avoidance in preventing bleeding episodes, as well as the perception of a more encouraging social environment, from both parents and peers. Intention to perform on-demand treatment as recommended was associated with a stronger normative parental pressure and a higher level of perceived behavioral control.

Finally, in additional analysis, we performed a multiple regression analysis for each of the four studied health behaviors where all the predictors were entered in a single model to estimate the R-squared values. The variance of intention accounted for by attitude, subjective norm, and perceived control (according to R-squared values) was 54% for recommended physical activity intention, 68% for discouraged physical activity , 22% for prophylaxis, and 54% for on-demand treatment. This indicates that psychosocial factors of the theory of planned behavior were more powerful in explaining intention to adopt discouraged physical activity than other behaviors.

Discussion

Considering the important and irreversible consequences of nonadherence to treatment in hemophilia, it is important to understand what leads children and adolescents to follow recommendations. This is the first study that systematically explored factors of health behaviors in hemophilia using a behavioral theory approach. We found that different psychosocial factors accounted for an important part of the variance of intentions for each behavior, suggesting that different approaches should be used to influence adherence for each domain.

The main finding of this study is the high variance in the intention to avoid discouraged physical activity due to psychosocial factors (68%). Considering that this behavior may be one of the hardest to follow for young patients (lowest intention and attitude of all four behaviors with a moderate difference with other behaviors), it should represent a valuable target for psychosocial interventions. Variance in intention for discouraged physical activity was also higher than for other behaviors, suggesting that young people with hemophilia differ greatly in their commitment to avoid risky physical activity. On average, patients spent 1.5 days per week engaged in discouraged physical activity for 15 minutes or more. As two or more bleeds in a joint may cause irreversible damage, assessing all social factors involved in the desire to practice discouraged physical activity would be beneficial, including attitudes and perceived pressure from parents and peers (Gringeri, Ewenstein, & Reininger, 2014).

Subjective norms related to parents was significantly related to intention to avoid risky physical activity ($r = 0.543$, $p < 0.01$) and children perceived less pressure from parents to avoid risky physical activity than that for other favourable behaviors (with moderate differences with other behaviors). It must certainly be difficult for parents to restrict their child from activities practiced by other children. Parents may also want to compensate for the consequences of their child’s physical condition, or for the guilt they may experience related to the genetic transmission of the disease (Beeton, Neal, Watson, & Lee, 2007 ; Cho, Kang, Kim, Ji, & Song, 2008). These observations promote the need for interventions that help parents deal with their feelings about restrictions (Dutreil, Rice, Merritt, & Kuebler, 2011).

Unsurprisingly, as reflected by subjective norm results, the role of parents appeared to be important, explaining individual intentions of two out of four behaviors in our study. Research has shown that information parents communicate to their child can greatly differ from the information they get from professionals due to a variety of biases (Gregory et al., 2007). Our results suggest that parental erroneous beliefs or opinions could easily transfer to children and are probably a relevant target to enhance children's commitment to their treatment plan.

Consistent with previous research, we found that peers may have a greater effect on physical activity than on treatment adherence (La Greca, Bearman, & Moore, 2002). Thus, if disclosure to peers appears appropriate, emphasis could be on special needs for physical activity rather than on providing information about medical treatment. Professionals should routinely ask patients to what extent the disease interferes with friendships or peer relationships, especially if restrictions in physical activity may lead to a sense of social isolation (La Greca, 1990; La Greca et al., 2002).

Another important result was that general attitude was associated with the intention to adopt risky physical activity. As attitudes are global affective evaluations, this stresses the need to address motivational issues beyond the necessary step of informing and educating. Information interventions alone are known to have only a negligible effect in improving adherence in pediatrics (Kahana, Drotar, & Frazier, 2008). A wide range of efficient interventions have been developed to address values and concerns, and modify behaviors in pediatrics (Erickson, Gerstle, & Feldstein, 2005). Following our results, interventions for children with hemophilia could help them feel more comfortable with restrictions about physical activity as well as develop more positive views of appropriate activities.

Although the general attitude appeared to be positive for recommended physical activity, the children of our sample did not seem to fully realize its benefits, as shown by a median neutral score on the item assessing treatment efficacy in preventing joint bleed. This may reflect the fact that communication with patients is more focused on avoiding risky physical activity than on the benefits of recommended physical activity. As some children may not have had bleeding episodes in years, making it hard for them to realize the long-term consequences of their actions, it would be very useful to increase patients' knowledge on treatment efficacy in order to promote adherence (Schrijvers et al., 2013; Young, 2012). In that sense, strategies addressing communication on such “hot topics” within the family should be developed (Bérubé, Mouillard, Amesse, & Sultan, 2016a).

Finally, our limited results on perceived control should be interpreted with caution. Given that most of our participants still relied on their parents regarding behaviors relating to their illness, perceived control may not have been validly measured in the present study. Further research should therefore address this topic in samples composed of older children and adolescents who are more autonomous with their treatment (e.g., age range 14-18 years old).

Limitations

The results found in this study should be carefully interpreted. First, the sample size prevented us from relying on basic inferential statistics. Consequently, this limits the external validity of our findings. Second, the correlational design of this study prevents us from drawing causal conclusions between factors and intentions. Third, the limited size of some of the correlations may also be due to a lack of variability or ceiling effects. Fourth, we used 7-point scales based on the recommendations for constructing a theory of planned behavior questionnaire and to allow enough variance on single-item scales (Ajzen, 2011). However, some authors have proposed to use 5-point scales with children might have difficulty to understand the difference between the possibilities on a 7-point scales (Touchèque, Etienne, Missotten, & Dupuis, 2016). The present research should be considered as a preliminary step to identify promising factors of self-care behaviors to be further tested in larger samples. The results support the need to systematically investigate these psychosocial factors to identify targets for future interventions.

Conclusion

To conclude, this study found that several psychosocial factors were associated with intentions of young people with hemophilia to behave in accordance with recommendations of the health care team. We found that past behavior, general attitude and attitude related to bleeding prevention, and subjective norm related to parents and peers explained a large proportion of the intention to avoid risky physical activity. Further studies should therefore target physical activity and how choices of activities are made in young people with hemophilia. Given the differential associative patterns across behavior domains, future studies should address the different dimensions of health behaviors in hemophilia, without limiting their research to medical treatment adherence.

Implication for practice

Hemophilia is a rare bleeding disorder for which psychosocial studies are much needed considering its important impact in the daily life of patients and their family. Identification of predictive factors of self-care can help identify targets for future interventions. Several psychosocial factors were associated with intentions of young people with hemophilia to behave in accordance with recommendations of the health care team. We found that past behavior, general attitude and attitude related to bleeding prevention, and subjective norm related to parents and peers explained a large proportion of the intention to avoid risky physical activity. These results stress the importance of assessing those psychosocial factors when promoting appropriate self-care in children with hemophilia.

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Declaration of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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Table 1. Sample description.

Characteristics	Mean (SD), Range	n (%) (*n* = 24)
Age (years)	11.8 (3.3), 6-18	
Country of birth		
Canada		20 (83%)
Other		4 (17%)
Type of hemophilia		
A (severe)		21 (88%)
B (severe)		3 (13%)
Type of treatment		
Prophylaxis		20 (83%)
Immune tolerance		4 (17%)
Bleeding episodes (past year)	4.3 (6.3), 0-24	

Table 2. Description of means and standard deviations of children's intention, past behavior and psychosocial factors for four health behaviors

		Physical activity		Treatment[a]	
		Recommended	Discouraged	Prophylaxis	On-demand
Intention	(1-7 scale)[b]	6.13(1.33)	4.75(2.25)	6.60(0.75)	6.70(0.66)
Past behavior					
Practice[c] (Nb days)		4.04(1.93)	1.54(1.94)		
Nonadherence[d]				38.45(13.71)	35.95(10.92)
Attitude					
General	(1-7 scale)	5.92(0.93)	5.00(1.62)	6.05(1.54)	6.15(1.53)
Bleeding prevention	Benefits (1-7 scale)	4.46(2.06)	5.42(1.69)	6.50(0.69)	6.25(1.16)
	Importance (1-7 scale)	6.79(0.42)	6.79(0.42)	6.79(0.42)	6.79(0.42)
	Benefits x Importance[e]	30.50(14.56)	37.04(12.22)	43.90(5.73)	42.05(7.82)
Normative pressure					
Parents	Pressure (1-7 scale)	6.58(0.72)	5.54(1.79)	6.70(0.73)	6.70(0.73)
	Influence (1-7 scale)	4.75(1.75)	4.75(1.75)	5.95(1.32)	6.60(0.68)
	Pressure x Influence[e]	31.79(12.58)	26.96(14.94)	40.35(11.19)	44.50(7.58)
Peers	Pressure (1-7 scale)	5.21(1.74)	4.58(2.10)	5.78(1.22)	6.06(1.24)
	Influence (1-7 scale)	2.50(1.35)	2.50(1.35)	2.72(2.19)	3.44(2.00)
	Pressure x Influence[e]	12.58(7.73)	10.42(6.97)	16.11(15.06)	21.19(13.97)
Perceived control	(1-7 scale)	6.17(1.13)	4.75(2.05)	6.50(0.89)	6.45(0.89)

Note. *Intention*: Intention to follow the recommendations; *Attitude*: general: General positive or negative attitude towards the recommendations; *Attitudes: bleeding prevention*: Attitude regarding the efficacy of the recommendations in preventing joint bleed

[a] *n* = 20 for this analysis, patients with inhibitors excluded

[b] Likert scale 1 (lowest intention) to 7 (highest intention to follow recommendations)

[c] Number of days of practice in a typical week

[d] Score at VERITAS-Pro/ PRN from 24 (most adherent) to 120 (least adherent)

[e] Items multiplied to reflect the importance given by the person in this aspect

Table 3. Partial correlations of behavioral intentions with psychosocial factors controlled for age and past year bleeding

		Intention to follow recommendations			
		Physical activity		Treatment[a]	
		Recommended	Discouraged	Prophylaxis	On-demand
Past behavior[b]	Physical activity	0.34	0.49^{*}		
	Adherence[c]			0.24	-0.20
Attitude	General	0.05	0.71^{**}	0.16	0.08
	Bleeding prevention	-0.01	0.61^{**}	0.43	0.40
Normative pressure	Parents	0.34	0.54^{**}	0.28	0.68^{**}
	Peers	0.41^{t}	0.48^{*}	-0.04	0.13
Perceived control		0.00	0.17	-0.16	0.50^{*}

[a] $n = 20$ for this analysis, patients with inhibitors were excluded
[b] Degree at which patients followed the recommendations in the past (some item score have been reversed)
[c] Adherence: We used reversed scores at VERITAS-Pro–PRN
$^{*}p < .05$, $^{**}p < .01$, $^{t}p < 0.10$

Introduction à l’article 3

Le troisième article s’insérait aussi dans l’objectif général d’identifier des facteurs reliés à la motivation des jeunes à suivre les recommandations de l’équipe médicale. Dans l’article 3, nous nous sommes intéressés strictement aux recommandations liées à l’activité physique. La deuxième catégorie de facteurs étudiés dans la présente thèse a été tirée du modèle du sens commun, une théorie qui s’intéresse aux perceptions qu’ont les patients sur leur maladie et sur l’influence de ces perceptions sur leur façon de s’occuper de leur condition de santé. En effet, il serait difficile d’avoir un portrait juste de l’hémophilie et de son traitement chez les enfants et les adolescents qui en sont atteints sans bien définir comment ils perçoivent cette maladie et le rapport qu’ils entretiennent avec celle-ci. Cet article visait à répondre aux questions suivantes : les jeunes ont-ils une perception pessimiste et préoccupante de leur hémophilie ? Au contraire, se sentent-ils en contrôle et relativement en confiance face à l’hémophilie ? Ces perceptions ont-elles un impact sur leur motivation à suivre les recommandations ?

Le concept de perceptions de la maladie a été opérationnalisé dans le modèle du sens commun (MSC) (Leventhal, Meyer, & Nerenz, 1980). Ce modèle décrit le processus dynamique et complexe par lequel les patients se représentent « la menace » que peut représenter la maladie sur leur santé, leur estime d’eux-mêmes, leur vie, etc. Le modèle distingue deux types de représentations de cette menace qui s’inscrivent dans des processus en parallèle. D’un côté, il y aurait les représentations cognitives de la maladie qui comportent les éléments suivants : une identité (le nom de la maladie et les symptômes auxquels la personne associe celle-ci), une durée anticipée de la condition de santé, des conséquences perçues (physiques, cognitives et/ou sociales), une ou des causes perçues aux symptômes, ainsi qu’un degré de contrôlabilité perçu (en lien avec les capacités de l’individu ainsi que la capacité du traitement à guérir ou contrôler la condition). D’un autre côté, les individus formeraient également des représentations affectives face à leur maladie, c’est-à-dire qu’une certaine réponse émotionnelle s’activerait face à la maladie, comme de la peur et de l’inquiétude. Ces deux types de représentations détermineraient les actions potentielles qui seront mises en place par l’individu sous forme de plan d’action pour contrer la menace que représente la maladie et des processus de coping émotionnel seront aussi enclenchés pour réguler les émotions qui y sont associées (Leventhal, Phillips, & Burns, 2016). L’anxiété liée à la perception de la menace serait l’élément déclencheur du système d’autorégulation. Une fois les actions mises en place et les processus de coping déployés, il y aurait une évaluation des résultats obtenus (*appraisal*) qui pourrait résulter en une modification des perceptions elles-mêmes, du plan d'action ou des mécanismes de coping afin d'obtenir des résultats plus satisfaisants (voir figure du modèle complet à l'Annexe B). Ces processus pourraient être à la fois conscients et/ou inconscients chez les patients.

Le MSC fait partie des théories de l'autorégulation puisqu'il stipule que les individus régulent leurs comportements en fonction d'un réseau de processus qui interagissent de manière réciproque. Les différentes composantes s'interinfluencent afin d'atteindre un certain degré de cohérence entre elles. Par exemple, un adolescent atteint d'hémophilie qui fait l'expérience de chaleur et de picotement sur un membre peut reconnaître ces symptômes comme étant annonciateurs d'un saignement interne et entamer les démarches nécessaires avec ses parents pour traiter le saignement avant que celui-ci évolue. Sa réponse émotionnelle et comportementale sera donc en lien avec ses expériences passées et ses connaissances à propos des saignements et pourrait par exemple être modérée, ce qui lui permettrait d'administrer une injection sans trop s'inquiéter. Si son activation émotionnelle était trop grande et qu'il pensait que la présence de tels symptômes pourrait mener à une hospitalisation ou que ses parents pourraient de fâcher contre lui pour s'être blessé en pratiquant un sport interdit, il est plausible qu'il évite de leur en parler et cherche à réguler son anxiété autrement, en se distrayant en jouant à un jeu vidéo par exemple. Un autre patient pourrait tout simplement associer les mêmes symptômes à une piqure d'insecte ou à un coup de soleil et continuer à faire ses activités sans aucune inquiétude. Ainsi, la théorie stipule que les patients agissent selon une certaine logique et que cette logique interne peut grandement différer de celle des professionnels de la santé ou de leurs recommandations. Les auteurs de la théorie mettent également en garde les cliniciens qui désireraient modifier certaines perceptions de la maladie chez leurs patients pour les rendre plus conformes au modèle scientifique. En effet, modifier l'équilibre des croyances pourrait amener les patients à vivre une certaine anxiété si l'intervention n'inclut pas un plan d'action précis ou n'enseigne pas de nouvelles stratégies de régulation émotionnelle plus adaptées pour compenser celles qui le sont moins (Cameron & Leventhal, 2003).

Les perceptions de la maladie du MSC ont été liées à maintes reprises à l'adhésion au traitement pour diverses conditions de santé (Broadbent et al., 2015; Hagger & Orbell, 2003). Plus spécifiquement, des perceptions plus négatives sont souvent liées à une moins bonne adhésion au traitement et à d'autres comportements de santé moins adaptés (Broadbent et al., 2015; Law, Tolgyesi, & Howard, 2014; Mosleh & Almalik, 2016). Des interventions efficaces ont aussi été développées en utilisant le cadre de cette théorie (McAndrew et al., 2008).

L'objectif de ce troisième article était de décrire les perceptions de la maladie et d'étudier leur lien avec l'intention des patients de : 1) pratiquer une activité physique recommandée et 2) éviter de pratiquer une activité physique déconseillée. Pour ce faire, nous avons décrit l'activité physique des patients selon les profils d'activité physique, incluant leur pratique dans le passé et leur intention de pratiquer ces deux types d'activités dans le futur.

De façon cohérente avec la littérature, nous nous attendions à ce que les perceptions de la maladie soient plus pessimistes ou « menaçantes » chez les individus qui respectaient moins les recommandations, comparativement à ceux qui respectaient les recommandations.

Article 3

Illness perceptions and their relation to physical activity in children and adolescents with hemophilia

Sarah Bérubé[1,2], Claudine Amesse[2], Serge Sultan[1,2]*

[1] Université de Montréal
[2] Sainte-Justine University Health Centre

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Abstract

Background: Hemophilia is a rare bleeding disorder in which illness perceptions have seldom been studied. Illness perceptions are important in predicting patients' behavior. Due to the risk of bleeding and joint damage, healthcare professionals often discourage some physical activities. Those restrictions can be difficult to follow for pediatric patients. This study investigates the relationship between illness perceptions, physical activity behavior, and intentions amongst this population. Methods: Twenty-four 6-18-year-old patients with severe hemophilia completed the Brief Illness Perception Questionnaire. A questionnaire assessing their level of physical activity and intentions toward safe and higher-risk physical activity was also administered. Clinical and socio-demographic data were collected. Associations were studied using hierarchical clustering of physical activity patterns, and Mann-Whitney U comparisons between clusters. Results: Perceptions ranged from slightly to moderately threatening, and 20 to 30% of participants had highly threatening perceptions on Consequences, Identity, Concern, and Emotional response. The subgroup who engaged in more high-risk physical activity and had stronger intentions to engage in this type of activity also held more concerns and perceived more symptoms. Conclusion: Patients at risk of non-adherence to recommendations concerning physical activity have more threatening illness beliefs that could be addressed during specific interventions or routine appointments. Addressing illness beliefs could be an element to behavior change. Strong emotional responses to hemophilia also stresses the need for psychosocial support strategies.

Introduction

Hemophilia is a rare bleeding disorder in which the blood does not clot normally. Hemophilia A and B are the two most common forms and involve coagulation factors VIII and IX respectively. In severe hemophilia, the coagulation factor's concentration in the blood is less than 1%, which exposes these patients to a higher risk of internal bleeding, especially in the joints (Srivastava et al., 2013a). Bleeding episodes in the joints, if repeated, can lead to important and permanent joint damage (A Gringeri et al., 2014). Although hemophilia is not curable, bleeding episodes can be prevented by appropriate self-care. Infusions of the missing factor protein on a regular basis (prophylaxis) or when needed (on demand) are usually at the core of the treatment plan (Srivastava et al., 2013a). In addition to replacement therapy, the official guidelines of the World Federation of Hemophilia also encourage the regular practice of safe physical activity (PA) (Srivastava et al., 2013b). Being physically active helps prevent joint bleeds by strengthening the muscles around the joints, amongst other benefits. However, certain types of PA are strongly discouraged by healthcare professionals and they vary depending on the severity of the hemophilia, the level of joint health, and the strength and coordination of the patient. Sports involving contact and speed commonly fall into this category because they might cause bleeding in the joints (Cailly Howell & Patel, 2017; Zourikian, Jarock, & Mulder, 2010). Throughout this manuscript, PA that poses a higher risk of bleeding for a specific patient will be referred to as a high-risk activity, as determined by healthcare professionals.

Limitations in sports are a common concern for children and adolescents with hemophilia (Limperg et al., 2016). The practice of higher-risk activities is common in pediatric hemophilia (Ross, Goldenberg, Hund, & Manco-Johnson, 2009). Social pressures and the desire to be perceived as "normal" can lead patients to engage in sports that are discouraged by their healthcare team (Bérubé, Cloutier-Bergeron, Amesse, & Sultan, 2017b; Williams & Chapman, 2011). Indeed, children and adolescents with hemophilia usually practice more moderate- to higher-risk activities as compared to adults living with the same condition (Forsyth et al., 2014). Another issue arises when young patients avoid practicing PA of any kind, which also exposes them to a higher risk of joint bleeds (Nazzaro et al., 2006). Preventing joint bleeding is important as only two bleeding episodes in the same joint could cause permanent damage, such as a limitation in the range-of-motion of a joint and chronic pain. Moreover, bleeding episodes have a negative impact on the quality of life of pediatric patients (Bullinger & von Mackensen, 2003; A. v. Gringeri et al., 2004; Poon, Doctor, & Nichol, 2014; Shapiro et al., 2001).

According to the Common Sense Model of Self-regulation (CSM), the way patients view, understand, and feel about their illness (illness perceptions) are important factors in determining their motivation to follow their treatment plan (Leventhal et al., 1980). These factors will influence the way they cope with their condition and which actions they may take. Illness perceptions have long been studied in health psychology and have been shown to predict adherence, coping, and health outcomes in adult and pediatric patients (Broadbent et al., 2015; Hagger & Orbell, 2003; Law et al., 2014). Some children may have difficulty in realistically conceiving the illness and understanding why it is necessary to follow recommendations of their healthcare team due to their level of cognitive development (Bir & Podmore, 1990). In fact, children and adolescents have been found to be more focused on current symptoms, short-term treatment gains, and more immediate consequences than adults (Law et al., 2014).

Illness perceptions are made of different components: identity beliefs (the extent to which the patient attributes symptoms to the illness), causal beliefs (the perceived cause of illness), timeline beliefs (the perception of the likely duration of the illness), beliefs about personal control and treatment control (whether the illness can be cured or controlled by one's action and by treatment), consequences (perceived consequences on one's life), understanding (the level of understanding of the illness), and emotional representations (emotional reactions to the health threat such as concerns, fear, anger, and distress) (Broadbent, Petrie, Main, & Weinman, 2006; Cameron & Leventhal, 2003). The relation between illness perceptions and self-management have been investigated in several studies in adults as well as in a pediatric patients (Broadbent, Donkin, & Stroh, 2011; Law et al., 2014). According to the model, the perception of a health threat will activate self-regulation processes (Cameron & Leventhal, 2003). Concordantly, some studies have found that stronger illness perceptions were associated with better adherence (Ross, Walker, & MacLeod, 2004; van Os, Troop, Sullivan, & Hart, 2017). However, according to the theory, when negative emotion arousal is too high or the health threat is perceived as unmanageable, affect regulation could become primary and lead to defensive denial vs to the management of the objective threat (e.g., through adherence) (Cameron & Leventhal, 2003). In the literature, more threatening beliefs are often linked to poorer health outcomes, poorer adherence to a health regimen, and lack of self-care behaviors, including PA in adults, children and adolescents (Broadbent et al., 2011; Broadbent et al., 2015; Law et al., 2014; Mosleh & Almalik, 2016; Skinner et al., 2003). Studies that focused on exercise found that more exercise was associated with a higher perception of control as well as a better understanding of the disease (Broadbent et al., 2011; Mosleh & Almalik, 2016; Skinner et al., 2003). In hemophilia studies, having less concern was associated with better adherence to treatment in adolescent and adult patients (Lamiani et al., 2015; van Os et al., 2017). Some contradictions were found: having a stronger emotional reaction (e.g., fear and anger) and a stronger perception of chronicity were associated with both a better and a worse adherence to treatment in two studies (Lamiani et al., 2015; van Os et al., 2017). None of those two studies focused exclusively on pediatric patients. The contradiction in the direction of the association could be explained by the use of different versions of the questionnaire, and differences in the level of perceived emotions and chronicity, as well as in health status, age, and treatment regimen (Lamiani et al., 2015; van Os et al., 2017).

Illness perceptions may play a key role in motivation for safe practice of PA in children and adolescents with hemophilia. Paying a closer look at them would allow us to better understand the experience of pediatric patients and, consequently, help practitioners develop targeted interventions to modify perceptions and influence behaviors. Very few studies either directly investigate younger patients' point of view on their hemophilia or focus on PA's psychosocial determinants. To our knowledge, no study has used illness perceptions to understand PA with this population. This is all the more important as interventions aiming at changing illness perceptions have led to significant outcomes in patient health and wellbeing, and have been shown efficient in changing self-care behaviors such as exercise (Broadbent et al., 2015; Jones et al., 2016; Keogh et al., 2011; Petrie & Broadbent, 2003; Petrie, Cameron, Ellis, Buick, & Weinman, 2002).

In this study, we first wanted to describe illness perceptions in children and adolescents with hemophilia, and, second, to examine the relationship between illness perceptions and usual practice of PA, and between illness perceptions and intentions to practice PA in the future (since it has been shown to be a proximal predictor of actual behavior) (Armitage & Conner, 2001). In order to do so, we wanted to determine if there were consistent profiles in our sample with respect to how children behave and think about recommended and high-risk PA, and investigate whether illness perceptions would differ across these profiles. Based on studies on the relationship between PA and illness perceptions in other illnesses, as well as studies on other self-care behaviors, we expected that illness perceptions in general would differ across profiles of PA, i.e., that more negative illness perceptions would be associated with poorer adherence to PA recommendations.

Materials and methods

Participants

Patients aged 6-18 years with severe hemophilia A or B (VIII / IX < 1%), treated at Sainte-Justine University Health Center and accompanied by one of their parents, were approached for this study which consisted of a one-time assessment during their appointment at the hemophilia clinic. All 31 patients who were on the list of the local hemostasis center and those who met the inclusion and exclusion criteria were approached to participate in this study. We excluded young patients who were diagnosed with a psychiatric disorder or had received their diagnosis less than a year before the study and were still adapting to the illness, in order to avoid outliers and collect answers from a relatively homogenous sample representative of the majority of young people with hemophilia. Of the 31 patients contacted for the study, 26 patients (84%) and their parents accepted to participate (Table 1). Reasons for participation refusals were due to not having a scheduled appointment at the clinic (*n* = 3) and lack of time (*n* = 2). Two participants were excluded from our study because of comorbidities (intellectual disability and severe attention deficit disorder), resulting in a final sample size of 24 pediatric patients with a mean age of 11.8 ± 3.3 years. Contacting all of the eligible patients respected the natural variation in participants' age, hemophilia type, treatment plan, etc. All of them spoke French. Questions were read to patients and a research assistant collected the answers. Young children were asked if they understood the questions before answering and all participants were encouraged to ask questions at any time during data collection. Children were assured that their answers would be kept confidential and they all, as well as their parents, gave their written informed consent before the study. The Sainte-Justine UHC Research Ethics Board approved this project. In this study, we used the same sample as in a previous study (Bérubé, Cloutier-Bergeron, Amesse, & Sultan, 2017).

Measures

To assess illness perceptions, we used the Brief-Illness Perception Questionnaire (Brief-IPQ), a short version of the Illness Perception Questionnaire-Revised (Broadbent, Petrie, et al., 2006; Moss-Morris et al., 2002). This questionnaire contains eight questions, each rated on a 11-

point Likert scale assessing illness perceptions of the CSM, as described in the introduction section. Questions go as follows: How much does your illness affect your life? (Consequences); How long do you think your illness will continue? (Timeline); How much control do you feel you have over your illness? (Personal control); How much do you think your treatment can help your illness? (Treatment control); How much do you experience symptoms from your illness? (Identity); How concerned are you about your illness? (Concern); How well do you feel you understand your illness? (Understanding); How much does your illness affect you emotionally? (e.g., does it make you angry, scared, upset or depressed?) (Emotional response). The last and open-ended question about the believed causes of the illness was not included, as hemophilia is a genetic condition generally diagnosed early in life. The Brief-IPQ is a measure with highly adequate psychometric properties. A meta-analysis concluded that this instrument is suitable for a range of conditions and has good concurrent and predictive validity (Broadbent et al., 2015).

To assess behaviors regarding PA, we first used an open-ended question inquiring which sports had been specifically recommended and discouraged to patients by their healthcare team. Then, participants had to report their PA level in a typical week for both types of activities (safe and high-risk). In this article, high-risk PA is defined as any physical activity that has been identified as posing a high risk of bleeding for the patient by healthcare professionals and therefore should be avoided according to one's treatment plan. These activities might differ between participants. Furthermore, considering that activities vary greatly according to the Canadian seasons, we inquired for both winter and summer activities and averaged both sets of results. We used the similar wording used in the Godin Leisure-Time questionnaire (Godin & Shephard, 1985): *In a typical week, how many days per week do you engage in this type of activity for more than 15 minutes?* Since the estimation of PA is not reliable for children under 10 years old, parents' estimations were used for these participants (Kohl et al., 2000).

Intentions to adopt future behaviors were also assessed as a measure of patients' motivation to follow recommendations in the future. We used one item for each type of PA (recommended or non-recommended). Following standard practice, the participant responded on a 7-point Likert scale, for example: *In the future, I intend to practice recommended/discouraged PA, 1-strongly disagree to 7-strongly agree* (Ajzen, 2006).

Statistical analysis

For the scoring of the Brief-IPQ and descriptive statistics, we reversed scores for items 3, 4, and 7 as indicated in the scoring instructions (Broadbent, 2010).

In order to describe participants' illness perceptions, we compared the mean score of our sample to the midpoint of 5 on the 11-point scale with a Wilcoxon signed-rank test to check whether responses were in the lower or upper half of the possible scale responses, representing respectively a lower perceived health threat (a more benign illness) or a higher perceived health threat (Broadbent, 2006).

To identify homogeneous groups of participants according to their tendency to follow or not to follow the recommendations around physical activity, we used hierarchical clustering (Ward, squared Euclidean distance) (Beaulieu-Prévost, Ouellette, & Achille, 2005). This produced agglomerative clusters of participants who were similar on the selected internal variables. The internal variables used for clustering were the average number of days of safe and high-risk PA per week as well as intentions to practice safe and high-risk PA in the future. The final number of clusters was determined by the study of the dendrogram and through the verification of the clusters' clinical interpretation (Mooi & Sarstedt, 2011). We compared illness perceptions across PA clusters using Mann-Whitney U tests. Effect sizes of the differences (*r)* were also calculated and interpreted according to Cohen's convention of small ($r = 0.1$), medium ($r = 0.3$), and large ($r = 0.5$) effect sizes (Cohen, 1992).

Results

Table 2 shows participants' mean scores for each of the illness perceptions of the Brief-IPQ (for median scores and interquartile ranges, see supplementary file 2). Low scores represent rather positive beliefs and high scores show the more threatening ones. Participants agreed that the illness was chronic (high mean for Timeline and small SD). According to the distribution of responses in Figure 1, two participants did not perceive their illness as being chronic. Participants' scores for Treatment control and Understanding were all equal or inferior to the midpoint of 5, thus in the portion of the scale representing a lower perceived threat. Mean score for Personal control was also under the midpoint of 5, although a few participants had higher scores (Wilcoxon, $p < .01$). This suggests that, in general, patients perceived having a relatively good control over their illness, believed that their treatment was helpful, and that they had a good understanding of their hemophilia. However, Consequences, Identity, Concern, and Emotional response were rated as representing a higher threat for participants. For these items, participants rated these perceptions as being moderately strong, i.e., they perceived a moderate level of consequence and symptoms, and experienced a moderate level of concern and other negative emotions in relation to their illness. There was a significant variability in answers to these aspects and about 20 to 35% of the sample answered 7 or more on the 0-10-point scale, with higher scores representing more threatening beliefs.

We conducted the hierarchical clustering procedure to explore individual profiles toward PA. Upon graphical examination of this dendrogram, we evaluated the greatest distance in which observations were combined (Mooi & Sarstedt, 2011). The most obvious solution when using this method was to use two clusters. These clusters were also clinically relevant since they represented two types of distinct attitudes toward high-risk PA: avoiding risk vs. engaging in it. Other clustering alternatives were not interpretable clinically and were thus discarded (see supplementary file 1). To characterize the two profiles, we compared participants' levels on internal variables and found that they differed on their past practice of riskier PA and intentions to engage in these activities in the future (see Table 3). Patients in Profile 1 practiced more high-risk PA (2.6 vs. 0.6 days per week) and had stronger intentions to engage in high-risk PA in the future than Profile 2 (5.1 vs. 1.7). Consequently, Profile 1 was labeled as the Risk Profile and Profile 2 as the Safe Profile. The two profiles did not significantly differ both on typical practice of safe PA and on intentions to practice these types of activities in the future. Further analysis also showed that the two profiles did not significantly differ in age (12.0(3.2) years vs. 11.7(3.6) years) nor in the number of bleeding episodes in the last year (3.8(6.3) episodes vs. 4.7(6.7)).

While exploring differences in illness perceptions across these profiles, we found that patients from the Risk Profile had significantly more threatening views of the illness overall (total score). They also reported more symptoms (stronger Identity), and were more concerned about their illness (stronger Concern) compared to their Safe Profile counterparts. Importantly, the size of these differences ranged from moderate to large (Cohen, 1992). We also performed further analysis to verify that the removal of the 4 participants that were on immune-tolerance therapy would not affect the present results. This was not the case with the exception of Concern, for which the difference did not reach significance ($p = .53$) when comparing the two profiles. It is unclear whether this difference in our results is due to individual differences, systematic differences between individuals with or without immune tolerance therapy, or a reduction in statistical power.

Discussion

In a study exploring illness perceptions and patterns of PA in 24 children and adolescents with hemophilia, we found that most dimensions of illness perceptions ranged from slightly to moderately threatening, with the exception of Timeline. Moderately strong perceptions were found in relation to Consequences, Identity, Concern, and Emotional responses. Moreover, there was a high variability in answers to these aspects and a significant proportion of participants (20 to 35%) estimated these negative perceptions as being very strong (7 or higher on the 11-point scale). On the other hand, participants were all rather positively confident regarding Treatment control and Understanding. Personal control was also high for most participants. We found consistent profiles describing behaviors and intentions toward PA in this sample, with the Risk Profile practicing more high-risk (discouraged by their healthcare professionals) PA and expressing stronger intentions to practice those activities in the future compared to the second group (Safe Profile). However, there was no significant difference in practice or intentions for safe PA. As for the associations between illness perceptions and individual profiles toward PA, participants in the Risk Group had significantly more threatening perceptions of their illness globally. They also perceived more symptoms of their illness and had more concerns related to hemophilia.

Our results highlight that, even though pediatric patients may be confident in the efficacy of their treatment and in their own capacity to control their condition, some aspects of hemophilia and its treatment can be particularly difficult for them on an emotional level. Physical dysfunctions, pain, the unpredictable nature of the illness, social challenges, and efforts to conceal their difference can all represent important sources of concern for pediatric patients with hemophilia (Nazzaro et al., 2006; Williams & Chapman, 2011). A large proportion of participants (35% or more) in our study expressed strong emotional representations of illness (7 or more on an 11-point scale), which is concerning considering that stronger emotional representations have been associated with depression, anxiety, and a worse quality of life in patients with diabetes, as reported in a meta-analysis on the Brief-IPQ (Broadbent et al., 2015). However, participants had an unanimously high level of Treatment control and Understanding, and a high level of Personal control, suggesting that healthcare providers and caregivers are sufficiently supportive in providing information about hemophilia and teaching children and adolescents with hemophilia about their medical treatment. Interestingly, these representations were similar between the Risk Profile and the Safe Profile. This could mean that youngsters who are inclined to engage in high-risk PA do not particularly feel the need to gain more information or to increase their self-care skills. Thus, they might not be interested in formal information-based interventions. Similarly, a meta-analysis on interventions for changing PA behaviors in children has concluded that education alone is unlikely to change behavior (Brown et al., 2016). However, some results also stress the need to objectively assess the degree of knowledge of patients, as some misconceptions can indeed be present and impede adherence to recommendations. The fact that a subsample in our study did not see hemophilia as a chronic illness was surprising. When exploring possible explanations for this phenomenon, we found a study on illness perceptions suggesting that adolescents and young adults with hemophilia have a tendency to see hemophilia as being less chronic than older adults do (Lamiani et al., 2015). However, younger age may not completely explain this misconception, as it may also be due to a lack of knowledge about the illness, which is independent of age. This explanation is supported by a study that has shown that patients of all ages, as well as caregivers, can lack basic and important knowledge about hemophilia and its treatment (Novais, Duclos, Varin, Lopez, & Chamouard, 2016). Furthermore, it would be important not to discard the possibility that these patients believed in an eventual cure considering the important advances in gene therapy (Herzog, 2016).

Despite the same average number of bleeding episodes in the last year in the two profiles, participants in the Risk Profile perceived more symptoms and were more concerned about their illness than those in the Safe Profile. Apart from the possibility that they experience more symptoms other than bleeding, e.g., chronic pain, a possible explanation is that children and adolescents in the Risk Profile were more vigilant concerning their hemophilia symptoms and were more concerned about their illness because they feared the negative consequences of their behaviors. This is consistent with the widespread approach of health professionals (including in our own center) that strongly emphasizes risks resulting from inappropriate PA. This would mean that freeing themselves from the constraints of their PA does not make them feel better in the long run. An alternative explanation lies in the use of denial by patients who perceive more symptoms. A meta-analysis that explored the relation between illness perception and coping behaviors found moderate to strong correlations between identity beliefs (perceiving more symptoms) and the use of avoidance/denial defined as the cognitive or behavioral attempts to ignore or avoid the existence of the problem or illness (Hagger & Orbell, 2003). This means that it is possible that children and adolescents with hemophilia who perceive many severe symptoms and are more concerned consciously or unconsciously choose not to adhere to recommendations in order to avoid the anxiety that comes with recognizing the presence of a chronic illness. Studies in other clinical populations have suggested that a moderate level of worry or fear arousal is optimal for a patient's engagement in adequate health behaviors (Phillips, Green, & Morrissey, 2012; Strong & Dubas, 1993). In adults with hemophilia, not accepting one's hemophilia has been recognized as a barrier to illness management, as self-care acts as a reminder of the illness (Schrijvers et al., 2015). There was also a trend for participants in the Risk Profile to perceive having less control over their illness and to experience more negative emotions. It is possible that the size of our sample was too small to reach statistical significance. Finally, the fact that there was no difference between the two profiles in terms of the number of bleeding episodes is somewhat counterintuitive, as one would expect that engaging in more high-risk PA would impact the number of bleeding episodes. A possible explanation resides in the total amount of PA. Participants in the Risk Profile tended to practice more PA overall when both recommended and high-risk activities were cumulated, benefits, such as stronger muscles around the joints,thus compensating for some risks of injury. It is also possible that participants' behaviors had not been going on for long enough to yield an impact on joint bleeds in our sample.

If these results were to be confirmed in large-scale studies, they should be translated into recommendations for clinical practice. In fact, our study suggests that a subgroup of patients is particularly at risk of non-adherence to recommendations concerning PA and that this group has more threatening illness beliefs that could be addressed during specific interventions or routine appointments. According to this study, pediatric patients who engage in riskier PA experience more concern and perceive more symptoms regarding their illness. This is contrary to a misconception one could have that patients who do not follow the recommendations do not realize the risks involved in their behaviors. Therefore, a strategy to promote safer PA could be to assess the concerns and negative aspects they already experience rather than putting emphasis on communicating the risks associated with high-risk PA. This idea is in line with motivational interviewing techniques that use and amplify adolescents' and adults' own motivation for change in order to create behavioral change (Strait, McQuillin, Smith, & Englund, 2012). For children of all ages, acknowledging and being sensitive to their perspective and feelings is part of an autonomy-supportive approach, which has been shown to influence health behaviors (Bérubé, Mouillard, Amesse, & Sultan, 2016b). Even though quality of life has been generally good in young patients with hemophilia since the introduction of home treatment, especially for adolescents on prophylaxis, a high percentage of our sample still reported having a strong emotional response to hemophilia, which stresses the need for psychosocial support strategies (A. Gringeri et al., 2004). Illness perceptions are rarely assessed by healthcare providers but individuals are generally open to discuss them (Petrie & Weinman, 2006). An interesting approach for discussing illness perceptions has been through peer group discussions, which allow children to express their inaccurate perceptions about the illness in order to then reframe them (Williams & Binnie, 2002). Other creative ways have been successfully used to assess illness perceptions such as the use of drawings (Broadbent, Ellis, Gamble, & Petrie, 2006).

It is important to recognize the limitations of this research. First, as hemophilia is a rare disease, our sample is limited and not randomly selected. Therefore, it is vulnerable to a lack of statistical power as well as selection bias, which may threaten external validity. Importantly, the study took place in Canada, where prophylaxis access is possible through the public healthcare insurance system. This decreases the financial burden and limits some of the stressors that might be present in families living with a child with hemophilia in other countries. Equally important, the correlational design of this study prevents us from drawing causal inferences between illness perceptions and PA patterns. Furthermore, our study included pediatric patients of a wide age range and our small sample size prevented us from studying differences between age groups. We recognized that patients from different ages have their own challenges often related to their developmental stage, and that the present study does not account for these specific differences. In addition, using parents' estimations of PA for children 10 years old and under while using self-report for patients over 10 can represent a risk of bias in data. Also, PA practice and illness beliefs were not objectively assessed, and self-reports may have been influenced by social desirability. While the Brief-IPQ has been successfully used with children from the age of eight, it has not been validated for patients that young (Broadbent et al., 2015). Pediatric studies on antecedents of self-care behaviors are extremely rare, all the more so in hemophilia. Future studies should address these limitations by using longitudinal designs and more extensive measures of self-care including PA. Since PA patterns acquired in childhood and adolescence tend to persist into adulthood, understanding early PA determinants and its prevention are paramount (Hirvensalo & Lintunen, 2011; Tercyak, 2007).

To conclude, in a study of 24 pediatric patients with hemophilia, we surveyed illness perceptions, patterns of behaviors, and intentions toward PA. We found that children had slightly to moderately threatening illness beliefs and that a significant proportion had highly threatening beliefs. Furthermore, we found that children who practiced riskier PA and had the intention to do so in the future also felt more concerned about their illness and experienced their illness as more symptomatic than the rest of the sample. Future studies are needed to confirm these results and to investigate the interplay of motivation and behavior in this population.

Authors' contribution

S. Berube and S. Sultan were responsible for the design of the project and for writing of the article. S. Berube was responsible of the recruitment, data collection, literature review and analysis. C. Amesse helped in recruiting participants, clinical advice and reviewing the literature. All authors approved the final version of the manuscript.

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Declaration of interest

The authors declared no conflict of interest.

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Table 1. Sample description (*n* = 24)

Characteristics	Mean (SD), Range	n (%)
Age (years)	11.8 (3.3), 6-18	
Country of birth		
Canada		20 (83%)
Other		4 (17%)
Type of hemophilia		
A (severe)		21 (88%)
B (severe)		3 (13%)
Type of treatment		
Prophylaxis		20 (83%)
Immune tolerance		4 (17%)
Bleeding episodes (past year)	4.3 (6.3), 0-24[a]	

[a] In our sample, 4 participants did not experience any bleeding in the last year

Table 2. Mean, standard deviation, and range on items of the Brief-IPQ for all participants (*n* = 24), Risk Profile (*n* = 11), and Safe Profile (*n* = 13)

Items	Description	Scale anchors	All	Risk Profile	Safe Profile	Effect size (*r*)
1	Consequences	none to severe	4.75(3.00), 0-10	5.45(3.33), 0-10	4.15(2.67), 0-10	0.24
2	Timeline	very short-term to forever	9.13(2.05), 3-10	8.91(2.21), 3-10	9.31(1.97), 3-10	0.14
3	Personal control[a]	excellent to no control	2.88(2.80), 0-10	4.00(2.97), 0-10	1.92(2.36), 0-8	0.38
4	Treatment control[a]	excellent to no control	1.00(1.44), 0-5	1.27(1.27), 0-3	0.77(1.59), 0-5	0.31
5	Identity	none to many severe symptoms	4.33(2.87), 0-9	5.82(1.99), 3-9	3.08(2.96), 0-8	0.47*
6	Concern	none to extreme	5.04(2.79)[b], 0-10	6.70(2.36)[c], 4-10	3.77(2.45), 0-7	0.51*
7	Understanding[a]	very clear to not at all	2.08(1.82), 0-5	2.64(1.86), 0-5	1.62(1.71), 0-5	0.28
8	Emotional response	none to extreme negative emotions	4.79(3.54), 0-10	6.18(3.28), 0-10	3.62(3.43), 0-9	0.36
Total	Threatening views (global score)		34.13(12.77[b], 4-55	41.80(9.82)[c], 25-55	28.23(11.83), 4-46	0.52*

All items are rated on a 10-point Likert scale

[a] Item score is reversed-coded, with higher scores representing more threatening illness beliefs

[b] *n* = 23

[c] *n* = 10

Difference between groups (Mann Whitney U): *p < .05, **p <.01

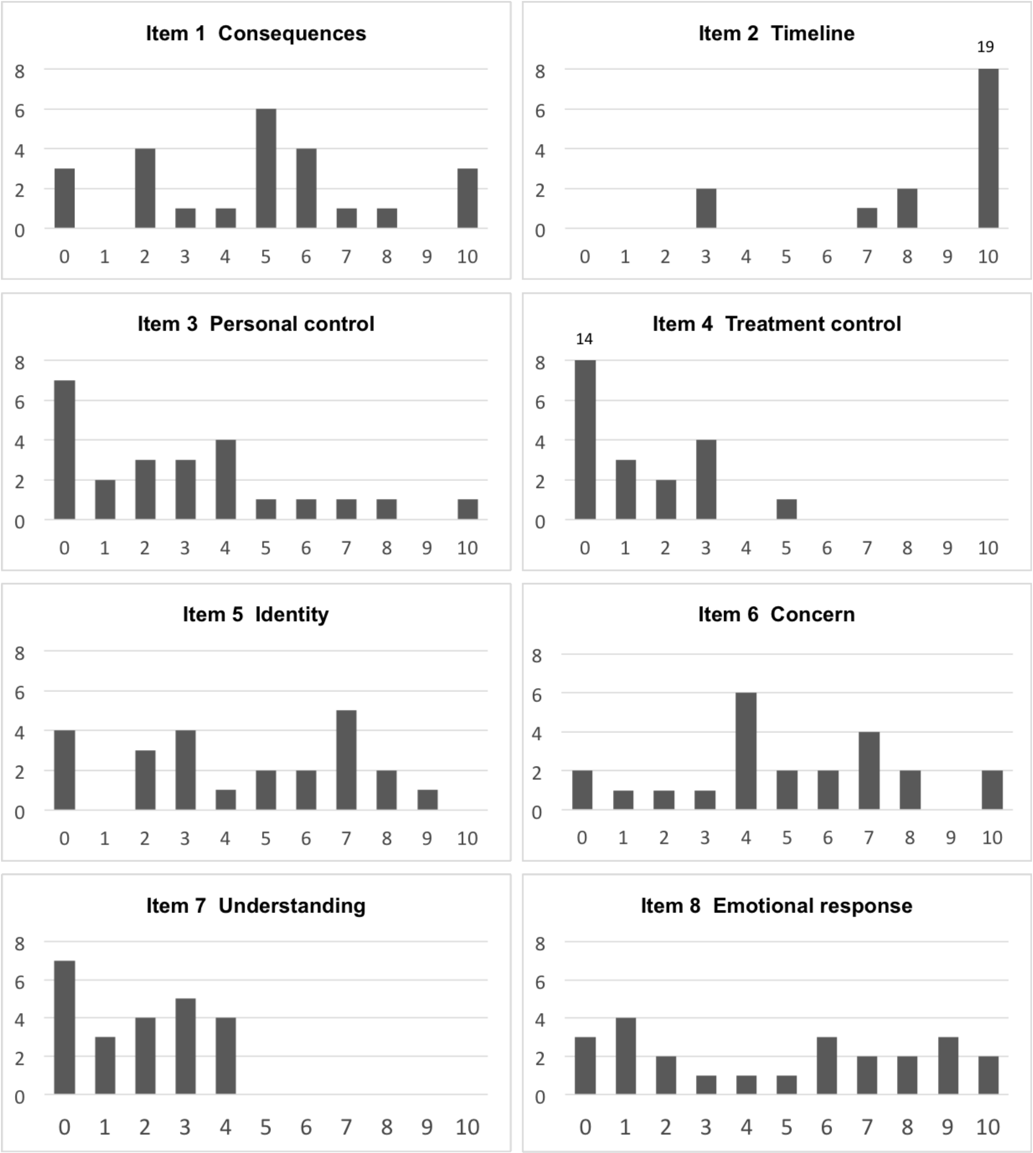
Table 3. Characterization of physical activity (PA) profiles in a group of 24 young patients with hemophilia (mean and standard variation)

	Risk Profile	Safe Profile	Effect size (*r*)
Physical activity practice[a]			
Recommended	3.59 (1.87)	4.42 (1.98)	0.22
Discouraged	2.64 (2.34)	0.62 (0.79)	0.51*
Intention to practice PA			
Recommended	6.09 (0.83)	6.15 (1.68)	0.22
Discouraged	5.09 (1.45)	1.69 (1.49)	0.75**

[a] Number of days of self-reported practice in a typical week (average score for winter and summer season)
Difference between groups (Mann Whitney U): $^{*}p < .05$, $^{**}p < .01$

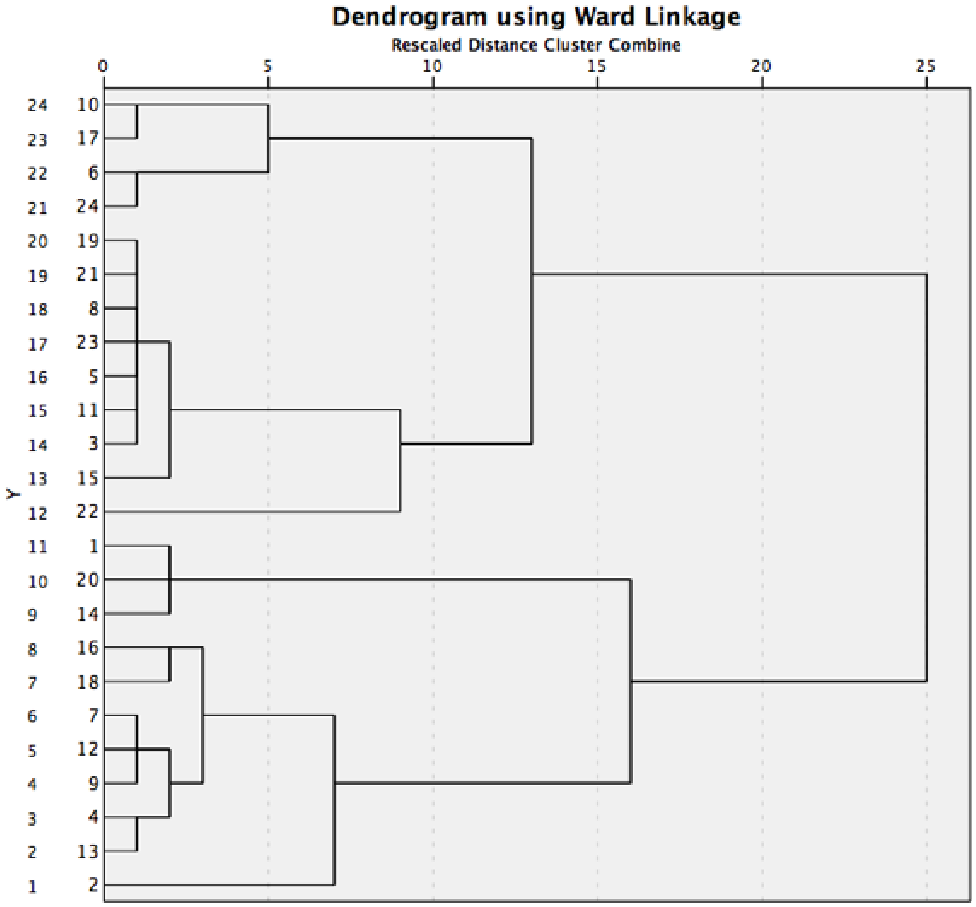
Note: We used hierarchical clustering (Ward, squared Euclidean distance). The internal variables entered for clustering were: average number of days of safe PA per week, average number of days of riskier PA per week, intention to practice safe PA in the future, and intention to practice riskier PA. This produced two agglomerative clusters of participants with similarities on the selected internal variables. Differences in mean scores for the two groups on the internal variables are presented in this table.

Figure 1. Frequency of participants' answers (vertical axis) for each scale value (horizontal axis) on items of the Brief Illness Perception Questionnaire (Brief-IPQ)



Supplementary file 1

Figure S1. Dendrogram resulting of the cluster analysis of high-risk and safe PA behaviors and intention using Ward's method.



Supplementary file 2.

Table S2. Median response (IQR) on items of the Brief-IPQ for all participants (*n* = 24), Risk Profile (*n* = 11) and Safe Profile (*n* = 13)

Items	**Description**	**Scale anchors**	All	Risk Profile	Safe Profile
1	Consequences	none to severe	5.0(4.0)	6.0(4.0)	5.0(3.5)
2	Timeline	very short-term to forever	10.0(0.0)	10.0(2.0)	10.0(0.0)
3	Personal control[a]	excellent to no control	2.5(4.0)	4.0(4.0)	1.0(3.5)
4	Treatment control[a]	excellent to no control	0.0(2.0)	1.0(3.0)	0.0(1.0)
5	Identity	none to many severe symptoms	4.5(5.0)	6.0(3.0)	2.0(6.5)
6	Concern	none to extreme	5.0(3.0)[b]	7.0(4.5)[c]	4.0(4.5)
7	Understanding[a]	very clear to not at all	2.0(3.0)	3.0(4.0)	1.0(3.0)
8	Emotional response	none to extreme negative emotions	5.5(7.0)	7.0(6.0)	2.0(6.0)
Total	Threatening views (global score)		34.0(18.0)[b]	42.5(17.3)[c]	32.0(16.5)

All items are on a 0-10 scale

[a] Item score is reversed in order for all higher scores to represent more threatening illness beliefs.

[b] *n* = 23

[c] *n* = 10

Introduction à l’article 4 et 5

Finalement, considérant les enjeux en hémophilie décrits dans cette thèse, il est apparu nécessaire de développer des outils d’intervention pour faciliter la prise en charge de la maladie dans la population pédiatrique. Dans les articles 2 et 3, nous avons vu que certains facteurs étaient liés à la motivation des jeunes à suivre leur plan de traitement comme recommandé. Cependant, les modèles dont sont tirés ces facteurs, c’est-à-dire la théorie du comportement planifié et le modèle du sens commun ne fournissent pas d’indications sur des moyens concrets pour les influencer (Ogden, 2014; Sniehotta, Presseau, & Araújo-Soares, 2014). Dans la théorie du comportement planifié, on indique qu’on doit essayer de changer les perceptions biaisées qui contribuent à une attitude plus négative, un sentiment de contrôle plus bas et à une norme subjective encourageant des comportements à risque (Sniehotta et al., 2014). Dans le modèle du sens commun, on indique que les interventions efficaces sont celles qui permettent une autorégulation efficace, en misant à la fois sur des stratégies centrées sur le problème ou sur les symptômes, à la fois sur des stratégies de régulation émotionnelle. Aucune précision n’est cependant donnée par les auteurs sur la façon d’y arriver (Cameron & Leventhal, 2003).

En découvrant la théorie de l’autodétermination en cours de projet, il nous est apparu évident que celle-ci était toute désignée pour pallier cette lacune. Cette théorie n’a pas été développée spécifiquement pour le domaine de la santé et de l’activité physique, mais elle a fait ses preuves dans ce contexte, notamment en permettant de prédire l’adhésion au traitement et l’activité physique (Deci & Ryan, 2004). En faisant une lecture critique des écrits sur la théorie de l’autodétermination, nous avons constaté que non seulement celle-ci peut servir de modèle conceptuel en soi pour l’intervention, c’est-à-dire avec ses propres facteurs, ses propres cibles, pour favoriser une motivation à long terme chez les patients, mais elle peut également servir de cadre directeur pour le développement d’intervention visant à modifier d’autres facteurs, provenant d’autres théories. En effet, cette théorie n’entre pas en contradiction avec les modèles théoriques utilisés dans les articles 2 et 3, comme nous en discuterons davantage dans la discussion générale de cette thèse. Au contraire, elle décrit des paramètres importants à considérer dans toute intervention psychosociale.

La théorie de l’autodétermination se concentre sur la motivation humaine, en particulier sur les comportements autodéterminés, c’est-à-dire ceux qui émergent de la volonté propre de l'individu plutôt que d'une source de motivation externe (récompense, punition, etc.) (Deci & Ryan, 2012). Le degré de motivation le plus persistent serait la motivation dite « intégrée », c'est-à-dire, une motivation qui est basée sur les valeurs profondes de la personne. Dans ce type de motivation, le comportement en question n'entre pas en conflit avec d'autres buts importants pour la personne. Également, cette dernière ne tenterait pas consciemment ou inconsciemment d'obtenir, par le comportement, des récompenses ou d'éviter des punitions externes ou internes, comme de ressentir de la honte (Deci & Ryan, 2017). La théorie stipule qu'afin de développer cette forme de motivation, les individus doivent trouver réponse à trois besoins fondamentaux, soit le besoin de compétence, de réciprocité relationnelle et d'autonomie. Elle stipule également que les individus ont une tendance naturelle à intégrer l'information qui est transmise socialement. Par contre, ce processus d'intégration peut être entravé lorsque les trois besoins psychologiques fondamentaux ne sont pas satisfaits (Deci & Ryan, 2012).

Pour faire le pont avec les articles précédents, nous pouvons dire qu'il ne suffit pas de viser à modifier certains des facteurs de la théorie du comportement planifié ou du modèle du sens commun dans les interventions s'adressant aux patients, il faut aussi considérer que la manière dont nous influençons ces facteurs afin de s'assurer de répondre à leurs besoins psychologiques fondamentaux. Travailler dans cette optique favorise une réelle intégration des apprentissages, ainsi qu'un type de motivation chez les jeunes patients qui risque de persister, même lorsqu'ils seront plus autonomes et moins encadrés par leurs parents.

Par une approche théorique clinique, nous avons formulé dans l'article 4, la possibilité et l'intérêt d'utiliser la théorie de l'autodétermination pour favoriser l'adhésion aux recommandations entourant l'activité physique et le traitement par injection chez les jeunes atteints d'hémophilie. Puis, plus concrètement dans l'article 5, nous avons élaboré et étudié la faisabilité et l'acceptabilité d'un projet d'intervention sous forme de livret d'information pour les parents d'enfants atteints d'hémophilie. Ce livret d'information vise à favoriser l'adaptation des jeunes et de leurs parents à la maladie et augmenter l'adhésion aux recommandations entourant l'activité physique. Il a été démontré dans la littérature sur les interventions visant à favoriser l'adhésion au traitement chez les jeunes patients atteints de maladie chronique que l'efficacité des différents types d'intervention était variable. Dans une méta-analyse sur ce sujet, il est ressorti que les interventions qui se centraient uniquement sur la maladie et sur son traitement étaient en général peu efficaces et que les interventions dites comportementales, celles qui enseignaient des habiletés comme la résolution de problème et formaient les parents à ces techniques étaient, elles, les plus efficaces (Kahana, Drotar, & Frazier, 2008). Nous croyons qu’il est important de développer des interventions qui vont au-delà des aspects concrètement liés à la maladie et qui visent également à soutenir le bien-être global des patients et de leurs familles. La perspective des articles qui suivent est d’aider les familles à s’adapter à la maladie et à développer des habiletés leur permettant une plus grande autonomie au quotidien (« empowerment ») et une meilleure qualité de vie.

Article 4

Motivational techniques to improve self-care in hemophilia: The need to support autonomy in children

Sarah Bérubé[1,2], Florine Mouillard[2], Claudine Amesse[2], Serge Sultan[1,2]

[1] Université de Montréal, Québec, Canada
[2] CHU Sainte-Justine, Montréal, Québec, Canada

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Abstract

Background. In pediatric hemophilia, caregivers are facing unique challenges to adherence and self-care in children and adolescents with hemophilia. Hemophilia treatment requires adequate prophylaxis and on-demand treatment, as well as a clear behavioral strategy to limit risk taking in terms of physical exercise and diet. Medication adherence rates of hemophilia patients have been reported to decrease during late childhood and adolescence. In the developing child, moving safely from parent-care to self-care is one of the greatest challenges of integrative care within this domain. There is a clear need for initiatives designed to increase an individual's motivation for treatment and self-care activities.

Discussion. Among motivational approaches, the self-determination perspective offers a useful framework to explain how the transition to self-care can be facilitated. We discuss how motivation regarding hemophilia treatment may be increased through parental autonomy support and we offer examples of applied communication techniques to facilitate autonomy-supportive caregiving. Although it has not yet been tested in the context of hemophilia, these communication techniques could potentially help caregivers promote adherence and self-care in children.

Summary. Confronted by unique challenges to adherence and self-care, caregivers of children with hemophilia should move from an exclusive focus on illness-management education to an integrative strategy, including motivation-enhancing communication. The self-determination perspective provides important proximal objectives (e.g., autonomy support) to maintain optimal adherence in adolescents as they move from parent-care to self-care. Future research initiatives should address the practice of these communication techniques and evaluate them in the context of hemophilia.

keywords : hemophilia, motivation, adherence, self-care, intervention, psychosocial, self-determination

Background

Hemophilia is a rare genetic disorder that can lead to bleeding episodes in the joints and muscles, which can result in permanent damages. Current treatment requires adequate routine administration of the missing coagulation factor (prophylaxis) or the administration of infusions when bleeding symptoms occurs (episodic treatment). Without adequate adherence, people with hemophilia tend to experience more frequent and severe pain and bleeding episodes as well as a lower quality of life (García-Dasí et al., 2015; McLaughlin et al., 2014).

Research has consistently found that adherence to treatment tends to decrease during late childhood and adolescence in patients with other chronic health conditions (Taddeo et al., 2008a). The same tendency was observed in hemophilia where adherence to prophylaxis has been shown to decrease drastically during early adolescence, when self-administration usually starts. In a recent survey of healthcare providers in 147 treatment centers around the world, 90% of children with hemophilia aged 0-12 years had "high" or "very high" adherence to prophylaxis compared with only 54% for those aged 13-18 years (Geraghty et al., 2006b). Reported barriers to adherence recommendations included such reasons as "lack of time" or "forgetfulness" (De Moerloose et al., 2008; Geraghty et al., 2006b; Hacker et al., 2001; Nazzaro et al., 2006; Schrijvers et al., 2013). Moreover, 25% of young patients who were not "early treaters" when bleeding occurred, reported that they did not have the clotting agent in their possession at the time and 29% stated that they did not recognize the symptoms of the episode (Nazzaro et al., 2006). These challenges could be partly addressed by offering patients and their family education in illness management skills. However, it has been shown that information interventions alone only have a negligible effect in pediatrics (Kahana et al., 2008). Clinicians also often recognize that these reported barriers are linked to motivational factors. Hemophilia patients may have only experienced a few bleeding episodes in their childhood due to adequate adherence to prophylaxis, as they were being treated by their parents. As adolescents, they may not realize the importance of preventive measures. This could lead to reduced adherence to prophylaxis recommendations or stopping their infusions (Young, 2012). Adolescents are also often described as being more present-oriented. Thus they may not be willing or capable of considering long-term consequences and may even ignore their disease because of their desire to be perceived as "normal" (Taddeo et al., 2008a). Adolescents may also be less inclined to listen to their parents and might want to test their limits (Taddeo et al., 2008a). Teenage rebellion, or the desire to free oneself from imposed constraints, has been described by nurses as a key factor in hemophilia nonadherence (Geraghty et al., 2006b). Motivational factors appear central to adherence and self-care behaviors so that patients do not see their treatment plan as being imposed on them by their caregivers.

Although fewer studies are available on other self-care behaviors than on adherence to medical treatment, it is probable that the same motivational factors influence adolescents' choices regarding physical activity and weight control. It has been shown that fostering motivation in patients can lead to better adherence to the recommended exercise (Chan, Lonsdale, Ho, Yung, & Chan, 2009). The World Federation of Hemophilia guidelines recommend appropriate physical activity to help promote fitness, neuromuscular development, coordination and healthy body weight in order to prevent joint damage (Srivastava et al., 2013a). Non-contact sports such as swimming will be encouraged, while high contact/collision or high-speed sports such as football or hockey will not be recommended for these patients (Srivastava et al., 2013a). Surprisingly and contrasting with the aforementioned guidelines, a national survey conducted in 2006 found that 60% of 459 young patients across the US managed their hemophilia by simply avoiding physical activity altogether (Nazzaro et al., 2006). On the other hand, a higher proportion than expected had engaged in high-impact activities in the last year (Cassie Ross et al., 2009). In a 2007 survey, 74 young patients with hemophilia in Germany were asked to indicate their primary motivations for participating in sports activities. “Having fun” or, “social aspects” were always chosen as the top two reasons for their participation, with “health benefits” consistently coming in third place (Fromme et al., 2007).

Adherence issues have been observed in hemophilia as in many other chronic conditions, despite important advances made in education management training (Duncan et al., 2012; Geraghty et al., 2006b; Jonasson, Carlsen, Sodal, Jonasson, & Mowinckel, 1999; Morris et al., 1997). This observation underlines the role of other important factors, such as motivation, when confronting adherence issues. As evidenced in more prevalent conditions such as diabetes, intervening on motivation and autonomy aspects may be highly effective in increasing adherence and self-care in pediatrics (Austin, Guay, Senécal, Fernet, & Nouwen, 2013; Austin, Senécal, Guay, & Nouwen, 2011). Parents also need to receive sufficient support in order to help their child accept and adapt to their illness and its management. In the present article, we explain how parents and caregivers can implement motivational techniques to help their child or adolescent gradually and safely move towards having greater independence. We offer practical communication strategies, which are likely to change motivation and thus increase adherence to the treatment regimen.

Discussion

Motivation for self-care

Various reasons, attitudes and beliefs may lead young patients to avoid general treatment recommendations, especially when they start self-infusing and making their own choices. Therefore, children and adolescents must be guided in developing and maintaining motivation for their treatment and health behaviors. This motivation should persist while parental supervision progressively decreases and the child starts self-infusing, which usually occurs before the age of 15 (Lindvall et al., 2006). Parents may understand the importance of following the recommendations and may become engaged in the management of their child's illness in the very early stages, but this motivation also needs to be transferred to the child as he/she gains more autonomy. It can be difficult for parents to find a balance between prohibiting behaviors and encouraging their child's autonomy and freedom of choice (Markova, Macdonald, & Forbes, 1980). Overprotection as well can have detrimental psychosocial effects on children (Cimarolli, Reinhardt, & Horowitz, 2006; Holmbeck et al., 2002; Janssens, Oldehinkel, & Rosmalen, 2009; Mak, 1994). However, overindulgence is also associated with negative psychological effects, especially in the context of hemophilia where children need to avoid potentially dangerous behaviors (Eiser, 2012). Few programs formally help parents deal with everyday problems related to hemophilia management (Dutreil et al., 2011). Typically, little information is usually given to parents to help them adequately address illness-related issues with their children, such as how to decrease the burden of injections or how to discuss risky sports involvement. If not addressed properly in early childhood, these issues may be perceived by children as restrictions to their autonomy, which could result in a lack of motivation to pursue appropriate behaviors when they are older.

Self-determination in children

One approach psychologists have used to understand and promote motivation is called Self-Determination Theory (SDT) (Deci & Ryan, 2002). According to this approach, it is possible to set limits without precluding children from becoming inherently motivated for their behavior and thus motivated to participate in activities that are not necessarily pleasant, such as infusions (Koestner, Ryan, Bernieri, & Holt, 1984). According to this theory, people have a natural tendency to internalize socially transmitted values, but this internalization process can be supported or hindered by their environment (Grolnick, Deci, & Ryan, 1997). This process may be viewed on a continuum ranging from being motivated by external factors (external regulation) to being motivated by internal factors (integrated regulation) (Ryan & Deci, 2000). A child would be motivated by external factors if he/she engages in the recommended activities in order to obtain parental approval, rewards or to avoid guilt (Ryan & Deci, 2000). This form of motivation has sometimes been associated with short-term benefits, but is often accompanied with anxiety and dissatisfaction and does not typically persist over time (Hagger & Chatzisarantis, 2009).

From a self-care perspective, it is desirable that young patients with hemophilia develop a more integrated form of regulation for their behaviors (e.g., engaging in physical activity because physical activity is important for one's health and is in line with one's core values). This type of motivation is more likely to guide adolescents into making healthy choices in the absence of parental supervision. SDT is empirically based and has validated instruments to assess each of its constructs, which facilitates the development and evaluation of interventions (https://www.selfdeterminationtheory.org).

Compared to other frameworks, such as motivational interviewing or empowerment, techniques from SDT can be incorporated into family dynamics and healthcare practices as a way to communicate to children about their illness and its management, and thus are specifically applicable to the developing child. It is also possible to use these communication tools with children of all ages. The sets of communication techniques derived from SDT aim at fulfilling an individual's innate psychological needs for autonomy, competence and relatedness, which in turn can facilitate and foster a more integrated form of regulation (Ryan & Deci, 2000). Among these needs, autonomy would be essential in achieving a more integrated form of regulation and for this reason, many interventions are referred to as "autonomy-supportive" even though they also respond to needs for competence and relatedness (Ryan & Deci, 2000). SDT has been applied to chronic conditions requiring complex treatment regimens and was found to be effective for many outcomes, including adherence and physical activity (Deci & Ryan, 2002). A meta-analysis supported the development and implementation of interventions using this approach in patients of different ages (Hagger & Chatzisarantis, 2009). It concluded that autonomy-support from healthcare providers predicts patients' autonomy and satisfaction of needs, which in turn are important predictors of self-care and health outcomes (Hagger & Chatzisarantis, 2009). A study has found that autonomy-support provided by parents can promote PA in children (eg pedometer-determined PA level) (Rutten, Boen, & Seghers, 2013). Besides the favorable outcomes for health-related behaviors, SDT is known to have both short- and long-term impacts in a variety of areas, such as academic performance, psychological well-being and social adjustment (Grolnick & Ryan, 1989; Joussemet, Koestner, Lekes, & Landry, 2005; Joussemet, Landry, & Koestner, 2008a; Ryan, Deci, Grolnick, & La Guardia, 2006). On the contrary, a controlling family environment can undermine the development of an integrated form of regulation. Such control refers to the act of pressuring the child to think, feel or act differently using various techniques such as inducing guilt, withdrawing love or invalidating feelings (Joussemet et al., 2008a). Several reasons can lead parents to adopt such attitudes including anxiety and concerns about their child's health status (Grolnick, Gurland, DeCourcey, & Jacob, 2002).

Strategies to increase motivation in children with hemophilia

Autonomy-support should not be confused with permissiveness or neglect. Autonomy supportive environments typically include a clear structure for children in order to give them opportunities for self-achievement, all the while preventing them from engaging in risky behaviors (Wendy S. Grolnick, Deci & Ryan, 1997). Existing interventions have shown that professionals and parents can be trained to use an autonomy-supportive communication approach where the child is helped in meeting his or her innate psychological needs (Chatzisarantis & Hagger, 2009; Joussemet, Mageau, & Koestner, 2014). This may be achieved by encouraging personal choices, independent problem-solving, taking initiative, and by participation in shared decision-making. In such environments, the child's point of view is acknowledged as well as his/her ideas and feelings. (Deci & Ryan, 2002; Wendy S. Grolnick et al., 1997; Joussemet, Landry, & Koestner, 2008b; Reeve & Jang, 2006). However, this can represent a challenge with a child with hemophilia as the need to limit health risks strongly encourages caregivers to be more controlling than autonomy-supportive, thus emphasizing obedience or compliance. Unfortunately, with this latter type of approach, the child will generally be less adherent to his treatment or recommended self-care behaviors.

Examples of motivation-focused communication

In order to illustrate how these motivational principles may be applied in practice, we have gathered examples of autonomy-supportive communications that could be used with children with hemophilia (Table 1). We illustrate four communication principles based on SDT as applied to real-life situations, which could be potentially encountered by these parents. The fear that children may hurt themselves may lead distressed parents to become overly controlling and protective, hence potentially removing all opportunities for the child to find his/her own solutions to daily challenges. Motivation research suggests that these children do not only need education in order to adequately limit their risks, but they also need to develop the feeling that they are responsible for their own behaviors (Wu et al., 2014). Hence the structure given to children should be most effective if they can experience a sense of volition and choice. Importantly, research on self-determination suggests that any trained professional can communicate this way, and transfer these communication strategies to parents, either as part of routine clinical appointments or during more systematic interventions (Williams & Deci, 2001a; Williams, Gagné, Ryan, & Deci, 2002). Long-term management of hemophilia could greatly benefit from the development and the evaluation of such an approach.

Conclusions

In summary, recent research has shown it can be difficult for children with hemophilia and their parents to implement the recommendations of the healthcare team into their daily life. Challenges are even greater during the transition from parent-care to self-care. Parents' systematic use of various communication strategies to help promote autonomy and appropriate self-care in young patients can be beneficial. Such strategies can help parents promote autonomy in their child while maintaining necessary boundaries. As illustrated in Table 1, the self-determination approach provides tools to help promote self-motivation in children with hemophilia during the transition towards self-care. We believe that understanding and using these communication principles can help caregivers to better address the motivation challenges in self-care faced by children and adolescents with hemophilia. Future research initiatives should address the development of standardized caregiver training following these principles and offer appropriate strategy to evaluate it.

Competing interests

The authors state that they have no interests, which may be perceived as posing conflict or bias.

Authors' contribution

SB wrote the manuscript.

FM designed an earlier version of a parenting skills intervention and wrote an initial draft.

CA gave clinical advice and edited the manuscript.

SS supervised the elaboration and wrote the manuscript.

Authors' information

SB is currently leading a research project as part of her Ph.D, which investigates the determinants of self-care in pediatric hemophilia. SB, CA, and SS are working on pretesting a standardized training initiative designed for professionals and parents confronted with pediatric hemophilia. If you are interested or wish to participate, please contact SB or SS for further details.

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Table 1. Application of autonomy support concepts to parenting a child with hemophilia

Topic 1: Acknowledging and being sensitive to his/her perspective, feelings and ideas Example: *The child comes back from school crying because his teacher said he/she could not play soccer during recess time.*			
Autonomy supportive		Non-autonomy-supportive	
1. Name the child's feeling. Acknowledge that it can be difficult.	*“You must have felt really angry when the teacher said you could not play soccer.”*	Being judgmental about the child’s feelings and ideas. Rationalizing or minimizing emotions.	*“Stop acting like a baby, you know why, you know the teacher did that for your own good, you should be grateful.”*
2. Show that you are listening and let the child find his/her own solution	*“I see… Hmm Hmm…”*	Interrupting Criticizing	*“We have told you many times that you can’t play, you should not even start to play.”*
3. Encourage the child to propose ideas and write them down, even the ones that are not suitable with his/her health condition.	*“Let's make a list of all the sports that you would like to play.”*	Making judgments about what he or she values as good or important	*“Health should be your priority.”*
4. Take into consideration his/her opinion about the suitability of the behavior.	*“Let’s see which ones are possible, or not, for you to play and why.“*	Imposing your opinion.	*“I enrolled you in swimming classes.”*
5. Ask questions to find out what the child likes in this specific unsuitable sport? Help him/her to be specific when they do not like something.	*“What do you like in this sport? Is it being with your friends?”* *“Why don’t you like swimming?”*	Trying to convince.	“*We are lucky that we have a pool right next to our house, many kids would be happy to have that*.”
6. Help the child to find alternatives that meet these interests.	*“Let’s see what could make these activities safe for you. Do you have any ideas?”* “*Which other sport would make you go fast like when people play hockey?”*	Impose solutions	*“Next time, you should explain to your friend that you can’t play.”*
Topic 2: Providing choices, minimizing control and involving the child as much as possible Example: *The child does not want to receive his injection in the morning.*			
Autonomy supportive		Non-autonomy-supportive	
1. As much as possible, give the child choices related to the management of hemophilia.	*“Would you prefer to watch TV during your injection or read a book ?”*	Imposing decisions, applying pressure or arguing	*“The doctor said you have to receive your injection in the morning, if you do not I am going to tell him and he won’t be happy.”*
2. Engage the child as much as possible in his treatment	*“Do you want to disinfect your skin while I prepare your injection?”*	Being inconsistent or too permissive	“*It’s ok I give up, we will do your injection tomorrow.”*
Topic 3: Providing structure and explaining the rationale when choices are not possible Example: The child hurts himself/herself playing outside and did not tell anyone, which caused a bleeding episode.			
Autonomy supportive		Non-autonomy-supportive	
1. Explain in a language adapted to the child's level of comprehension as to why the preventive behaviors are important.	“*You have to call mommy so we can inject the little soldiers in your blood that will fix the boo boo.”*	Giving too much information at once, accentuating long-term consequences or scaring the child.	“ *If you don’t receive your injection, you might not be able to walk when you are my age.”*
2. Set important limits for the child and stay consistent.	*“You always have to call me when you feel in pain and I will come and take care of your infusion.”*	Setting excessive rules.	*“You have to call me before engaging in any physical activity.”*

3. Encourage questions. Encourage the child's ideas and for them to look for answers.	*“Do you know why your knee is getting bigger? Why don’t we look up on the internet to find out or we can call the nurse tomorrow?”*	Avoiding discussion	“*We have talked about it many times, you know what you have to do.”*
Topic 4: Showing compassion for the child and providing non-judgmental feedback			
Autonomy supportive		Non-autonomy-supportive	
1. Provide feedback that is not judgmental.	*“It’s a good thing that you called me, even though you felt ashamed that you played a sport you were not supposed to.”*	Categorizing the child.	*“You are irresponsible, I always have to check up on you.”*

Article 5

It's difficult to say no: Development of a parenting booklet about physical activity restrictions and recommendations in pediatric hemophilia

Sarah Bérubé[1,2*], David Ogez[1,2], Jennifer Aramideh[1,2], Claudine Amesse[2], Claude Julie Bourque[2],

Claire Longpré[2], Lorraine Muise[2], Ariane Levesque[1,2], Serge Sultan[1,2]

[1] Department of Psychology, University of Montreal, Quebec, Canada
[2] Sainte-Justine UHC, Montreal, Quebec, Canada
[3] Department of Pediatrics, Université de Montréal, Quebec, Canada

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Abstract

Physical activity restrictions can be particularly difficult for children with hemophilia and their parents, as most children want to engage in the same activities as their friends or siblings. This study aimed to assess parents' need for a booklet on how to manage physical activity restrictions based on the Self-Determination Theory and gather their suggestions for the booklet's improvement in a focus group. Data was analyzed using thematic analyses to highlight to what extent the needs of parents were met by the booklet, as well as their suggestions for improvement. A discussion on the methodology of the booklet's creation is also presented.

Introduction

Hemophilia is a bleeding disorder in which blood clotting is prolonged due to a missing clotting factor. Hemophilia patients require regular infusions of the missing clotting factor, as well as important precautions for the detection and prevention of bleeding episodes. Repeated muscle and joint bleeding can lead to arthropathy, chronic pain, and loss of range of motion in the long-term (McLaughlin et al., 2014; García-Dasí et al., 2015). After diagnosis, parents must quickly learn to handle many tasks, such as securing the home, recognizing bleeding symptoms, and learning how to infuse, in order to keep their children safe (Shaw and Riley, 2008). Parents often feel overwhelmed by their new responsibilities (Edmunds et al., 2007). They may also feel uncomfortable with administering painful infusions and they may worry about their child's safety (Beeton et al., 2007b).

Parental care evolves as the child ages. A new area of questions and concerns usually arises when the child becomes interested in organized sports. Although physical activity is highly recommended to prevent bleeding in the muscles and the joints, patients with severe hemophilia are commonly advised to avoid certain types of sports that are at risk of causing bleeding, such as football, and hockey (Mulder et al., 2004; Anderson and Forsyth, 2005; Broderick et al., 2012). Physical activity restrictions vary according to disease severity, joint health and muscle strength (Srivastava et al., 2013; Petrini and Seuser, 2009). Physical activity restrictions can be particularly difficult for children, as most want to engage in the same activities as their friends or siblings (Williams and Chapman, 2011). Children commonly react with sadness and anger when given physical activity restrictions. Balancing the child's desire for a normal active lifestyle and the need to protect them from risk represents a dilemma for parents (Shaw and Riley, 2008). Some decide to allow their children to participate in high-risk sports and hope that they will not injure themselves, while others tend to adopt overprotective behaviors (Beeton et al., 2007b; Abali et al., 2014). Non-compliance with physical activity recommendations issued by healthcare providers is common among children with hemophilia (Ross et al., 2009; Bérubé et al., 2017; Nazzaro et al., 2006).

The Self-Determination Theory (SDT) is helpful in understanding the process by which parents can set limits without compromising their child's well-being, in addition to increasing their self-motivation to comply with healthcare providers' recommendations (Deci and Ryan, 2002). According to this theory, parents can foster the development of a more autonomous and long-lasting form of motivation for health-related goals in their child by adopting an autonomy-supportive style of communication (Ryan and Deci, 2017). This theory is particularly helpful in the context of growing children who gradually need to acquire more autonomy in regard to their illness management. Studies on the SDT have shown that self-motivation for self-care health behaviors, even the ones that are not pleasant in themselves, can be fostered in children and adolescents. For example, parental autonomy support has been shown to preserve patient glucose monitoring frequency during adolescence, when glucose monitoring typically decreases (Wu et al., 2014). An environment that supports autonomy has also been shown to promote greater motivation for physical activity (Lim and Wang, 2009; Huéscar Hernández et al., 2020).

Autonomy-supportive parenting is focused on meeting the three basic psychological needs of children: autonomy, competence and relatedness. Meeting the need for autonomy involves the active nurturing of the child's capacities to self-regulate using techniques like taking the child's perspective, offering meaningful choices, and encouraging initiative and self-expression. Meeting the need for competence refers to using scaffolding strategies, providing non-evaluating feedback, and setting limits in a non-controlling way. Finally, meeting the need for relatedness refers to being involved with the child in a warm, caring, and non-judgmental manner (Deci and Ryan, 2002). Further information on the theoretical background of this study, as well as the SDT's communication strategies, has been published elsewhere (Bérubé et al., 2016).

Studies have highlighted the need to support parents of children with hemophilia in managing their responsibilities (Beeton et al., 2007b; Bottos et al., 2007). However, only a few studies on interventions for parents have been conducted in hemophilia and no known empirically tested interventions focus specifically on helping parents manage physical activity restrictions (Dutreil et al., 2011; Kennedy et al., 2017; Penica and Williams, 2008; Cuesta-Barriuso et al., 2014). A literature review confirmed that, when properly designed, health information leaflets can increase satisfaction, knowledge and adherence to treatment (Sustersic et al., 2017). Among other benefits, the availability of educational materials increases the effectiveness of verbal communication (Wizowski et al., 2008).

In the literature, the methodology used for the development of health-related or psychosocial information material for patients and their family is rarely detailed. Although guidelines are available, they remain broad and do not detail the concrete steps to accomplish the development and need assessment of information material using a mixed method (Oliver et al., 2008a; Farrell-Miller and Gentry, 1989). Furthermore, knowledge transfer has repeatedly been identified as an important issue in the scientific community. It is paramount to support patients and their family by providing them with access to quality information materiel derived from empirically validated models.

The aims of this study were:

1. To develop, with the help of healthcare professionals in hemophilia, a theory-based booklet for parents of children with hemophilia focused on how to manage physical activity restrictions and recommendations;
2. To assess parents' need for the type of material developed (i.e., a theory-based booklet) and to gather their suggestions for the booklet's improvement in a focus group;
3. To improve the booklet based on the suggestions elaborated in the focus groups with parents and meetings with the healthcare team.
4. To present a detailed methodology on the creation, adaptation and assessment of the need and acceptability of a theory-based booklet.

Methods

Booklet development

The methodology for the booklet development was based on recommendations for the production of written material for patients. (Oliver, Asselstine, Peterson, Rosati-White, & Stephenson, 2008; Sustersic et al., 2017). The booklet content was elaborated in collaboration with a nurse, a psychologist and a social worker from Sainte-Justine University Health Centre's Hematology Clinic, in Montreal, Quebec, Canada.

The healthcare professionals were first approached to discuss the need for providing parents with written material on physical activity management. The healthcare professionals expressed that there was a lack of written materials in hemophilia, particularly on physical activity management. At this first meeting, we held a brainstorming session to determine the target population, and the key concepts that should be addressed in this booklet. Following this meeting, a first version of the booklet's content was produced by the research team using the SDT and taking into account the healthcare professionals' ideas and key concepts. A second meeting with the healthcare professionals was held to review the booklet's content. They were invited to suggest ideas and changes. Their suggestions were immediately discussed to reach consensus within the healthcare team. Subsequently, each professional was met individually to work on the writing. A final meeting with all healthcare team members was held to achieve the version to be presented to participants. This version did not include images, although text boxes indicated where the images would subsequently be added. This was done with the idea that participants would feel more comfortable to criticize or propose major changes to a draft document than a document that looked in its final form. Further information on the adaptation of communication tools from the SDT to the specific context of hemophilia has been published elsewhere (Bérubé et al., 2016).

Booklet evaluation and suggestions for improvement

Participants

For this study, we approached parents of children with severe hemophilia treated at the Hematology Clinic. At the clinic, patients from the same age group have the same biannual appointment where they meet with each of the healthcare professionals during the day and participate in different group activities. We identified two groups representing an acceptable range of school-age children: 5-7 year-olds and 8-9 year-olds. An informative email was sent to all eligible parents and we contacted all parents who did not opt out after receiving the email. We aimed to recruit 5 to 10 parents for each of the two focus groups. Parents aged less than 18 years, and children or parents with a severe psychiatric disorder were excluded from this study. In total, eleven parents (eight mothers, three fathers) of pediatric patients aged 5-7 years old (60%) or 8-9 years old (40%) with hemophilia A (60%) or B (40%) were recruited and participated in the focus groups. In the 8-9 year-old group, at least one parent of every child that attended the hemophilia day at the clinic accepted to participate, while in the 5-7 year-old group, the parents of two children who attended the hemophilia day refused to participate in the study, saying they were not interested. The Sainte-Justine UHC Research Ethics Board approved this study (#2018-1877).

Procedure

The study design was a sequential mixed method based on a questionnaire, and focus group interviews about the booklet to evaluate its pertinence and acceptability (Creswell et al., 2003). First, parents participated in a 30-minute information session about the study during which the booklet was presented. They were then invited to fill out a questionnaire about the booklet to share their opinions on different dimensions (see Measures). Finally, they took part in the focus group interview, which placed emphasis on the elements that had been negatively evaluated in the questionnaire. Parents were invited to discuss each of these elements and to suggest ways to improve them. In total, there were two focus group interviews, one for each age group of children.

The interviews were designed and conducted according to the recognized principles of focus group interviewing (Wilkinson, 2015). The facilitators' role (SB, DO) was to ensure the fluidity of the discussion with open-ended questions. They were also responsible for ensuring that the discussion remained focused on the booklet and its improvement. If participants mentioned issues, concerns or needs that were out of the scope of the present study, they were told that time would be set aside at the end of the interview to discuss these aspects. Particular attention was given so that all parents were able to express their opinion. The interviews were audio recorded and transcribed verbatim.

Measures

We developed a questionnaire (See Supplementary File 1). Our questions were inspired by ones used in previous studies (Kazdin, 2005; Sahler et al., 2002; Ogez et al., 2019). The questionnaire included 36 items relative to 8 categories: the format (5 questions), the clarity of the information (2 questions), the structure (1 question), the length (2 questions), the expected positive effects (17 questions), the possible negative effects (2 questions), the usefulness and ease of implementation of the advice (4 questions), and the target population (3 questions). The responses were provided on a 6-point Likert scale for each suggestion, from 0 "strongly disagree" to 5 "strongly agree". For each category of questions, space was provided to allow participants to write comments.

Analysis

Descriptive analyses were led on the scales completed by the parents. We compared the mean score of our sample to the middle scale point of 2.5 on the 0-5 scale with a Wilcoxon signed-rank test, to indicate whether mean perceptions were positive, negative, or neutral in our sample. Qualitative data (textual responses to open-ended questions in the questionnaires) and interview transcripts were processed and encoded using the NVivo 11 software by analysts who received appropriate training specifically for this study. A descriptive thematic analysis method was used to create an inventory of themes by two independent coders (SB, DO) (Massey, 2011). Agreement was reached after two meetings to reach the final coding structure and code descriptions. Our analysis was based on articulated data, that is the information that was expressed by participants in response to the question posed (Massey, 2011). The objective was to ensure that all improvements proposed by the participants were identified, organized and grouped together to facilitate the editing stage. In presenting the results from the qualitative data, participants were identified by alphanumeric codes in the subsequent paragraphs (P1 to P11). Since the aim of the focus group was to obtain rather short and technical responses focused on improving the booklet, only quotes that significantly added to accuracy or understandability were presented in the results. We embedded short quotes in the text, and longer quotations were presented in a distinct paragraph.

Production of the Final Version

We met again with the psychologist, the nurse, and the social worker to critically analyze the central themes (or suggestions for improvement) to assess whether they considered that the suggested changes should be implemented. Since the number of parents was deliberately limited, each comment was considered to likely represent the opinion of a significant number of parents. The themes were thus considered as equally important whether they represented the opinion of one or several parents. Each theme (i.e., suggestion for improvement) was evaluated based on its usefulness, its feasibility in a booklet-type documentation, as well as the extent to which it met the aim of this specific booklet, current scientific knowledge, and the values of our healthcare center (e.g., fostering the empowerment of people and communities). We worked to obtain a consensus between healthcare professionals, as to whether the proposed changes should be adopted. When this consensus was reached, we met individually with the members of the healthcare team to modify the booklet's writing and presentation (the final version is available in Supplementary file 2).

Results

Booklet development

The consultation process with our hematology clinic's psychologist, nurse and social worker provided a rich brainstorming where many ideas were proposed. It was decided that the target population would be parents of school-aged children with hemophilia. The reasoning behind this decision was that it is generally at school age that children begin to compare themselves and want to move and play in the same way as their friends. While the healthcare team was not introduced to the Self-Determination Theory concepts prior to the initial meeting, their suggestions on parenting strategies were largely in line with them. This facilitated the production and edition of the booklet in collaboration with the healthcare team. Consensus was easily reached when one professional wanted to change, add or remove a topic. The only exception was that they could not easily decide who would be the resource person to refer parents to if they felt the need to talk or to have more information on the topic. It was finally decided that the message would be that every healthcare team member was there to provide support by listening to parents and/or referring them to the right specialist.

Booklet evaluation and suggestions for improvement

Responses to questionnaires

Each parent completed the survey before the focus groups commenced. Results are presented in Table 1. According to the mean scores compared to a neutral point of 2.5 on a 0-5 scale, participants generally agreed that the booklet was clear, well structured, and that the advice was useful and easy to use. Parents believed that the booklet was suitable for parents of children of all ages, but only moderately for adolescents. According to their answers, the format (booklet) was moderately appreciated. They also thought that the booklet could be longer and provide more daily tips. While they reported few expected negative effects, they perceived a moderate level of positive effects. Many changes were proposed by participants to increase the booklet's impact and usefulness during the focus-group interviews.

Qualitative analysis of parents' evaluation and recommendations from the focus groups

A) Format, length, and structure

During the focus groups, there was a consensus that the booklet was clear and easy to understand. Most parents considered booklet-type documentation to be a good and convenient format. P9 explained that a paper format allows parents to keep it for the time they will need it. However, P9 also felt that the booklet should be a little longer to give enough information without becoming too long to read. Importantly, some parents (P2, P3, P9) also expressed that the booklet was only an introduction and that they would need more help with the aspects discussed in the booklet.

When parents were asked what they thought about the way the information was structured, the majority responded that it was well divided. However, one parent (P2) thought the information should be given in bullet points rather than in a continuous text. Another parent (P1) suggested keeping the structure as it was, but to add a summary of the important information to remember at the end of the booklet, an idea that was well received by P2 and other parents. All parents also agreed that a list of resources, activities and links to more information on the subject would be useful if added at the end of the booklet.

B) Target population

Most parents suggested that the booklet should be distributed as soon as possible after the announcement of the diagnosis. Some parents (P9, P11) pointed out that parents, after a diagnosis like hemophilia, usually tend to search for information on the internet about the course of the illness and the limitations their children will have to face, making the booklet more useful during this period. One parent in the 8–9-year-old focus group (P3) said that parents are *masters of their trade* after their child reaches a certain age and that the booklet would not be useful to them. P1 discussed the importance of applying the communication tools from the booklet as early as possible with their child.

> *P1 : Well indeed I find that it is a booklet that should be given to you much earlier, because... precisely to build all this bond of trust there... and how we will interact with our child I find that it is from the beginning you know*

For P9, the content would be useful to some people and not to others depending on many factors: children's health status, treatment type, knowledge and parenting skills, etc.

C) Content

Some themes were highlighted by the parents as the most appreciated: the notion of teamwork with the child to find solutions (P2, P3, P9 would have liked more emphasis on this aspect), P11 liked to be reminded that help was available to them if they needed it. Other parents (P6 and P11) indicated that the most useful advice they retained from the booklet was to listen to their child rather than rushing to solutions.

> P6: *In that booklet, I make it a point, is that maybe to listen to him and let him talk instead of telling him you should do that. To say, how do you feel about it? What do you understand? To let him find his own solutions...*

P9 and P10 expressed that the booklet's possible comforting effect lied in its common sense which would reassure them that they are already doing the right thing for their child. However, P9 found that focusing on restrictions could generate some stress in the reader, and P10 said that it was important to dedramatize the illness. These parents (P9, P10) also expressed that the booklet was not specific enough to hemophilia and was lacking specific examples on how to apply the recommendations.

D) Missing content

Parents also highlighted different themes they felt were missing from the booklet. All agreed that mention should be made of how to encourage children to be physically active and how to valorize children. It was brought up that boys develop their self-esteem through their participation in sports (P2, P9, P10, P11). Therefore, some parents (P1, P10) thought the booklet should highlight that children who grow up with hemophilia acquire unique strengths: *They are courageous (...) they are resilient (...)*.

Some parents (P7, P9, P10) added that parental discourse should focus on possible and interesting alternatives. In this line, they (P10, P11) suggested adding a list of ideas for safe physical activities for people with hemophilia. The idea of integrating these activities early in their child's life also appeared fundamental to parents (P7, P9, P10). P11 added that it was a good opportunity for parents to try new sports with their child and to share a unique moment that promotes the relationship.

Many parents (P2, P3, P10, P12) mentioned that the notion of building a long-lasting trusting relationship was essential, since establishing a secure bond would help their child to integrate and refer to the rules. They also thought that this relationship might discourage the child from hiding injuries that would require attention. Many parents (P2, P3, P10, P11, P12) felt that they should empower their children in their decisions, such as talking to them about the risks that they face in certain sports. These parents (P2, P3, P9, P10, P12) also deemed important that the booklet more realistically address the notion of striking a balance between banning and allowing certain sports, well as promoting flexibility to encourage parents to refrain from using excessive protection.

Some parents (P2, P3) asked whether they should disclose their own emotions to their child. According to P3, expressing emotions to the child promotes a climate of cooperation. Many parents (P2, P3, P9, P10) mentioned that they themselves experienced a kind of grieving process when they realized that their child would not be able to play certain sports, and, therefore, experienced some of the same emotions as their child. Several parents (P2, P3, P4, P5) would have liked the booklet to mention that they have a right to feel guilty or sad and that it takes time to adapt to new challenges. P12 was concerned that restricting sports could cause depression in their child. Some parents (P10, P2, P3) would have found it helpful to have examples of specific questions to ask their child to help them express themselves.

All other propositions that were not directly linked to the booklet are briefly addressed in Supplementary File 3 to guide further research.

Production of the final version

When presented with the booklet evaluation results, healthcare professionals decided which suggestions of change should be applied to the booklet. The suggestions made by the parents and the decisions made by the healthcare team are summarized in Table 2.

Discussion

It is recognized that healthcare settings should not only meet the medical needs, but also the practical and emotional needs of patients and their family (Oates et al., 2000). In this study, we (1) developed a booklet intended for parents of children with hemophilia based on the SDT, (2) assessed parents' need for this type of material using a questionnaire and focus groups as well as (3) adapted the booklet according to most of their suggestions. Data collected helped us to refine the booklet in collaboration with healthcare providers.

The booklet development process with healthcare professionals went as planned. Having group meetings and one-on-one meetings with the staff made the writing process easier. Obtaining consensus on the points to be modified following parents' comments from the focus groups was done without difficulty since the clinical staff members had already established certain guidelines in their communication with the parents. The anonymity of the comments also seemed to allow the team to distance themselves from the criticism that may have been raised by the participants.

As in other studies, parents easily described the challenges related to their child's illness, and the strategies they used to try to overcome them (Williams and Chapman, 2011). The focus group allowed the team of researchers and healthcare providers to discover relevant themes to add to the booklet. Interestingly, many of the themes that parents felt were missing from the documents were in line with the SDT: building a trusting relationship, having a teamwork approach, finding alternatives (Bérubé et al., 2016; Ryan and Deci, 2017). Some parents sought to foster a relationship of trust to prevent their child from lying to them about accidents or engaging in risky physical activity. The ability to lie and to keep secrets is part of the healthy development of a child, but can also be a problematic behavior in some instances (Talwar and Crossman, 2011). In hemophilia, small secrets can have big consequences, e.g., the child hiding a bleeding accident or lying about engaging in a high-risk activity for fear of reprimand. It was decided that the booklet would explain the normalcy of occasional lies, but also promote, through parenting tips, a warm and sensitive relationship between the parent and the child, which could in turn reduce the number of lies. A study on lies in adolescence has shown a relation between parenting that supports autonomy and adolescents' identification to the honesty value as well as perceived low costs/high benefits of honesty, which ultimately predicted honesty (Bureau and Mageau, 2014).

Another important aspect that emerged from the focus groups was that parents had questions about their own emotions and sought ways to understand and cope with them. They sought reassurance but also wondered whether they should disclose some of their emotions to their child. A study on mothers of children with hemophilia aged 8 to 12 years showed that 21% of these mothers reported clinical distress (Limperg et al., 2016). Parents sometimes think that giving their child less information or hiding negative emotions may protect them in return, but studies on the subject seem to support the added value of true parental communication, within a certain limit (Karnilowicz et al., 2018; Claflin and Barbarin, 1991). Parents' emotional experience was also described as a grieving process in the focus groups. The grief of the healthy child is a well-known topic in the literature on parenting a child with a chronic illness (Kepreotes et al., 2010). In hemophilia, the grieving process surrounding physical activity restrictions is sometimes particularly difficult for fathers, as sports are culturally considered as an important aspect of fatherhood (Myrin Westesson et al., 2015). Parents in our study felt that the booklet was only an introduction and asked to receive a list of resources to accompany them, demonstrating their openness to receive more support. It is to be noted that at the hemophilia clinic, parents are invited to participate in different workshops and meet on a biannual basis: psychoeducational workshops for parents of children of different ages, groups and discussions with other parents, one-on-one meetings with the nurse, the doctor and the physiotherapist, optional meetings with the social worker and the psychologist, etc. Their interest in the booklet can be interpreted as a need to obtain information in different formats. The paper material can allow some parents to think more introspectively from their home and can also act as a reminder of the information that is explained in more detail during different activities.

Parents have conflicting decisions to make concerning their child's physical activity, as they often want to let their children engage in the same activities as their peers. This sometimes brings them to rationalize that higher-risk sports are sometimes acceptable (Beeton et al., 2007a). We observed variability in compliance with recommendations in our participants. While some of our participants were very adherent or partially adherent, others said that they did not impose any restriction over their child's physical activity, and this decision was sometimes hidden from the healthcare team. Some parents have expressed their discomfort with the emphasis on restrictions in the booklet and would have preferred that efforts be made to balance restrictions and permission. In fact, the idea of the booklet was specifically aimed at helping parents set necessary limits in a way that would not be detrimental to children despite the difficult emotions this might trigger. This can even have a positive impact on children's well-being and self-motivation according to studies on the SDT (Edmunds et al., 2007; Núñez and León, 2015). We modified the booklet to reflect the positive aspects of setting limits and tried to make the booklet version attractive to parents, even though the subject is often a sensitive one. Importantly, the booklet's purpose was also to encourage safe physical activity. An effort was made not to encourage overprotection which could have harmful effects on the child's physical and psychological health. There is a consensus in the literature on the importance of encouraging physical activity for people with hemophilia (Harris and Boggio, 2006; Negrier et al., 2013). Furthermore, physiotherapists and other health professionals can recommend adjustments to certain sports to make them safer (Harris and Boggio, 2006; Negrier et al., 2013).

While approaches for developing interventions and material vary greatly, guidelines usually recommend using a focus group to assess needs before creating any material or intervention (Oliver et al., 2008b; Moscoso et al., 2013). In this study, since the need for this type of documentation was clearly stated by the healthcare team and since we knew, from the literature, that there were many issues surrounding physical activity in children with hemophilia, we decided to present participants with a draft version of the booklet. We believe that this way of proceeding enriched the dialogue between parents during the focus groups and provided a concrete support for their discussion. It may be difficult for some parents to conceive what kind of information might be helpful to them without being presented with some ideas. Importantly, participants were also invited to express if this type of documentation corresponded to their needs. Further studies will assess the effects of using the booklet.

Some limitations must be considered when interpreting the results. This is a single-site study based in Canada, where prophylaxis access is possible through the public healthcare insurance system, which limits the results' generalizability. The intervention methods and the recommendations may differ substantially from one treatment center to another. Furthermore, the sample was conveniently selected based on dates of clinical appointment.

Conclusion

In conclusion, with the help of healthcare professionals, we conducted a mixed-method study to develop a booklet on managing physical activity restrictions and recommendations for parents. We also assessed parents' need for this kind of material, collected their suggestions for improvement, and reviewed the booklet according to their suggestions. The results showed that despite the high-quality support offered at the Hematology Clinic for children and their family, parents seem to need better support and information on the issues related to physical activity. Parents expressed they needed a booklet that would be reassuring and would normalize their emotions. Parents were also particularly interested in concrete and specific information, such as the importance of silently listening to their child instead of rushing to solutions. They also wanted more parenting tips such as ideas of questions to ask their child to better understand their perception of the illness and its impact on their life. The study's output is a booklet whose implementation could be studied in the hematology clinic. It is also ready to be adapted to other hemophilia treatment centers.

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Declaration of interest

The authors declared no conflict of interest

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Table 3. Parents' responses as to their opinion about the leaflet

Categories	M	SD	Min	Max	*p*	N of items	*N*
Format	3.11	1.29	0.00	4.60	0.11	5	11
Clarity of the information	4.59	0.54	3.50	5.00	0.00**	2	11
Structure	4.45	0.68	3.00	5.00	0.00**	1	11
Lenght	3.10	0.88	1.50	4.50	0.07	2	10
Envisaged positive effects	2.49	1.00	1.00	4.18	0.92	18	10
Envisaged negative effects	1.23	1.17	0.00	3.00	0.01*	2	11
Advices usefulness/ease of use	4.18	0.53	3.50	5.00	0.00**	4	11
Suitableness to age-groups							
5 years old and below	4.00	1.18	2.00	5.00	0.01*	1	11
between 6-12 years old	4.09	0.83	3.00	5.00	0.00*	1	11
between 12-17 years old	3.00	1.95	0.00	5.00	0.42	1	11

Note. Item's rating: 0 (totally disagree) to 5 (totally agree); *M* different than neutral point, * $p < .05$, ** $p < .01$.

Table 4 Summary of perceptions and suggestions for improvement of the leaflet according to the focus-group interviews

Theme	Parents' suggestions for improvement	Modifications / Decisions
1 - Format, structure	The leaflet is an introduction and parents need more help concerning these aspects	This will be considered in the clinic's practices.
	Replace text by bullet points or add a bullet point summary at the end	A bullet point summary was included at the end of the leaflet
	Add a list of resources for more information/help	A list was added at the end of the leaflet
2 – Target population	The leaflet should be handed as early as possible	It will be handed when the healthcare team addresses physical activity restrictions with the family
3 – Content	Might generate stress/should highlight more positive aspects of the illness rather than stressing on limitations	Positive outcomes were highlighted (e.g., that recognizing limitations is an opportunity to find solutions together and strengthen the bond)
	Information not specific enough to hemophilia	Specific examples and images were added
	More emphasis should be made on the idea of the child and the parent working together as a team	More emphasis was brought to the notion of togetherness
4 – Missing content		
Valorization	Should help parents valorize and encourage their child	This was added in the section on body image
Alternatives to activities	Should focus on the possible alternatives to high-risk activities (rather than limitations) and their implementation as early as possible in the child's life	It is important that the leaflet gives importance to both limitations (feeling of loss, disappointment), as well as on alternative activities. The notion of an early implementation in the child's life was added
Trusting relationship	Should address how to build a trusting relationship (so that it prevents the child from lying or hiding accidents)	Information about the lies in childhood and their function in the normal development was added, as well as parenting advice
Personal responsibility	Should help parents with empowering their child and making him/her more aware of the risks	The fact that young children cannot fully grasp the notion of risks was added and emphasis on parental responsibility was made clear
Permission vs restriction	Should encourage a healthy balance between permission and restriction	The leaflet aims to increase parents' capacity to tolerate sadness and to set limits to their child
Grief	Should address the process of grief for parents and patients	The word "grief" was avoided (too complex) The notion of loss/disappointment was added
Self-disclosure	Should encourage a healthy disclosure of parents' emotions to their child	Information about this was added

Parent’s involvement	Encourage parents to try new sports with their child	We have added a sentence to encourage parents to practice new activities with their children
Considering the future	Add clear information about typical course of illness and issues	This is out of the scope of the leaflet A section about parents’ emotions was added and aims to normalize certain concerns
Reassurance	Should reassure parents about the normality of their emotions	Information was added to reassure parents about the legitimacy of their own emotions
	Should give information on psychological issues children may and may not face	Information about the role of activity on psychological well-being was added. We also stressed the importance of a positive body image.
Ideas of questions	Add examples of questions to ask to children to be interested in their emotions/point of view	Examples were added

Supplementary file 1.

Evaluation "Accompanying your child living with hemophilia" - Questionnaire 1

In order to evaluate the material presented to you, could you complete the following questionnaire by placing your answer on the grid on the right: "0" meaning strongly disagree and "5" meaning strongly agree.

The information …

1) is clear and comprehensible
2) is well organized
3) is written in terms that are easy to understand
4) provides sufficient information

Personal remarks about the material:

The leaflet's advices…

5) are useful for everyday life
6) are well explained
7) are sufficient in number
8) are simple to apply
9) are easy to remember
10) are suitable for children 5 years old and under
11) are suitable for children 6 to 12 years old
12) are suitable for adolescents aged 12 to 17 years old

Personal remarks concerning the advices:

Effects of the information

If you had received the leaflet just before your child started school, it ...

13) would have brought you useful information
14) would have changed certain ways of doing things with your child

15) would have helped you to manage your child's tantrums
16) would have helped you to regain a sense of control
17) would have helped you to better understand your child's needs
18) would have promoted a better relationship with your child
19) would have promoted better communication with your child
20) would have made it easier to forbid your child from taking part in certain physical activities
21) would have a comforting effect on you ("vous aurait fait du bien")
22) would have reduced your stress
23) would have helped you to better control your reactions and emotions
24) would have promoted a better relationship with your health care team
25) would have helped you make a request for help if needed
26) would have helped your child to better adapt to the illness
27) would have had a beneficial effect on your child's mood
28) would have had a beneficial effect on your child's health
29) would have had beneficial effects for families other than your own
30) would have bothered you
31) would have generated concerns

Personal remarks about the leaflet's effects:

Format

About a paper format as it was presented to you…

32) would prompt you to read it
33) would prompt you to reread it
34) would prompt you to keep it
35) would prompt you to share it with others
36) This material has its place in the hospital

Personal remarks about the format:

We thank you warmly for your participation!!!

Supplementary file 2.

Final version of the booklet

ACCOMPANYING MY CHILD WITH HEMOPHILIA

INTRODUCTION.

Being a parent isn't always easy. Accompanying a child with a health condition such as hemophilia presents particular challenges.

Current medical knowledge stresses the importance of preventing bleeding into the joints to avoid long-term complications. Certain sports activities help make muscles stronger and reduce the risk of bleeding. Others, on the contrary, may increase the risk of joint bleeding. Your healthcare team can help you identify activities that are safe for your child.

Have you ever found it difficult to forbid your child from taking part in an activity? Has your child ever been sad or angry about not being allowed to participate in a physical activity that could cause bleeding? It is likely that you answered yes to one of these questions. These are difficult situations that parents of children with hemophilia may encounter.

This information leaflet proposes a few tips to help you communicate with your child about physical activity. You will be able to draw inspiration from it in your daily life and adapt it to your situation.

WELCOMING AND NAMING EMOTIONS

Children can experience all kinds of emotions related to their illness, whether they are pleasant or unpleasant. We all need to feel supported and welcomed in what we are experiencing. When you recognize that your child is experiencing a difficult emotion, it may be helpful to assist them in identifying it by expressing in words what you think the feeling is.

(ADD IMAGE: "What's going on? I feel like you're sad, is that it? ")

This response may seem obvious, but it is essential. In this type of exchange, children feel that someone is concerned about what is happening to them. Often, it is not necessary to go any further. Once the emotion is named, try to keep listening and see how your child reacts. Staying in silence with children helps them get in touch with and soothe their emotions. Your simple presence and your attention are comforting for your child.

(ADD IMAGE)

This way of accompanying children facilitates reflection and allows them to find their own solutions to problems they may face.

When this support is not enough, for example, in the presence of intense emotions, it may be necessary to give clear instructions to help your child calm down (e.g., ask your angry child to sit down, to take deep breaths).

(ADD IMAGE)

Worrying about your child is normal, especially when your child has a particular health condition. It should be kept in mind that all children will experience different emotions in their lives, some enjoyable, others less enjoyable. It is by letting children experience certain emotions, such as disappointment, that they learn to tolerate them and to face challenges.

If you have any doubts about your child's emotional well-being or development, talk to your healthcare team.

LISTEN TO YOURSELF AS WELL

As a parent, it is normal for you to experience all kinds of emotions. It is common for parents to feel disappointed, sad or angry to learn that their child will have certain physical activity restrictions. It can also be difficult to see your child sad or angry.

When you're experiencing a difficult emotion, it is not always possible to listen to your child or to make decisions. When this happens, it is appropriate to say: "I'll *think about it" or " what you're telling me is important, but right now I'm tired, we'll talk about it later".* By responding this way, you are expressing that you find your child's concerns important, and you are announcing that you will discuss the subject together when you are more disposed.

(ADD IMAGE)

Some parents worry that their child might see them sad or angry. What is most important in such a situation is for the child to feel like their parent is still taking care of their needs. For example, you can tell your child, "even if I cry, I can take care of you". It may be helpful to name the emotion you are feeling. It can be a good occasion to tell your child that it is normal to experience some unpleasant emotions from time to time. This often has a reassuring effect on children.

You may also feel overwhelmed by certain emotions. Just like your child, you need to be listened to and heard. Do you have anyone you trust that you can talk to?

(ADD IMAGE)

Taking care of your emotions will also have a positive impact on your child. When you feel well, it is easier to welcome another person's emotions.

FOSTER UNDERSTANDING

We all need to understand what's happening to us. When children understand why certain activities are not allowed and when they are accompanied in expressing their emotions, it is easier for them to adapt to the situation.

You may want to ask your child some questions to get a better idea of their understanding of the decisions that are being made. To avoid yes/no answers, it is always best to ask open-ended questions that encourage children to elaborate. For example, "What did you understand about what the nurse explained to you?". Finding answers together by talking to the healthcare team is always a win-win situation!

(ADD IMAGE)

It is also normal for young children to not understand everything about their illness. Since their thinking is mainly focused on the present, it is more difficult for them to fully grasp the future impact of their choices and to have long-term vision. Therefore, it is important for parents to make the final decision on which physical activities will be allowed and which ones will not.

ANTICIPATE DIFFICULT SITUATIONS

In the parents' absence, children sometimes have to make decisions that are not always easy, such as whether or not to participate in a physical activity. It is possible that your child may decide to participate in an unsafe activity without telling you. Often, children act in this way because of a desire to forget their illness, to test their physical capacities or to be like their friends.

No matter how you decide to approach this situation with your child, it is important to help them identify the emotions and concerns that may have pushed them to act this way. It may be useful to determine together how they could act when a similar situation arises again. Children sometimes need help to know what to say or do in difficult situations. Perhaps it might also be time to review which activities meet your child's new needs with your healthcare team?

BODY IMAGE

Children need to grow up with a positive image of their body. Moving during physical activities allows children to feel confident and helps them like their body, which is why it is essential to find activities that are fun and safe. When children discover activities that they enjoy at a young age, they take a liking for movement. They can also develop physical skills such as balance and coordination. Studies show that physical activity reduces anxiety and promotes good psychological health.

(ADD IMAGE)

You can ask children if they feel good about their bodies. This teaches them to listen to their body. Do they need to move? to stretch? to rest?

FEELING COMPETENT AND DEVELOPING AUTONOMY

Take a moment to inquire about your child’s favorite physical activities. For example, is it the speed that your child prefers, the competition or spending time with friends? These cues will allow you and your child to find a safe alternative that meets your child’s interests. Are there other sports that allow to go fast? Could your child be a referee or coach in a sport that captures their attention?

(ADD IMAGE)

Do research together. These moments help children feel heard and sense that their tastes and interests are important to you. And why not take part in the new activities with your child and enjoy some great family moments!

Allow your child to make choices as often as possible, for example by choosing the color of their equipment. This will provide your child with a sense of autonomy. Growing children need to feel like they are more and more capable of doing things on their own and of making decisions.

Creativity is your best ally!

IN SUMMARY

- Help your child identify and name their emotions.
- Cultivate silent listening! Your presence is sometimes sufficient and allows children to find their own solutions.
- Talk to someone you trust about your own emotions.
- Ask open-ended questions to check children's understanding of hemophilia and the recommendations that are made to them.
- Help your child find solutions and answers to difficult or embarrassing situations in advance.
- Ask questions to learn more about what your child thinks makes an activity interesting (speed, agility, the fact that it is a team sport, etc.).

- With your healthcare team, identify stimulating and safe activities that suit your child's taste and allows your child to move and stay healthy!

RESOURCES

Don't forget, your healthcare team is there to answer your questions. The pivot nurse, or any other member of the team, will be able to refer you to the resource that will best meet your needs. Together, we will be able to help your child integrate hemophilia harmoniously into his/her daily life!

(ADD LIST OF RESOURCES)

Supplementary File 3.

During the interviews, parents also expressed different needs and concerns that were out of the scope of the leaflet development. We dedicated time at the end of each focus group to discuss those issues, which could be addressed in future research. Here are the main issues which we make available for future references.

Firstly, many parents (P8, P9, P10, P11) thought that there should be some documentation addressed to the children directly to help them face the challenges around physical activity. They thought this could be part of the leaflet itself or in the form of a storybook like Kimo, a book that teaches children about hemophilia and infusions (Amesse, Longpré, Normandin, & Privé, 2014).

Several parents (P3, P4, P7, P9, P10) also expressed that it was difficult for them to find educative material and tools to hand out to teachers and school workers to explain the illness and all they have to know to prevent a bleeding episode.

P2 also proposed that there should be tools available to help them deal with their child’s emotions in their daily life such as a list of emotions, a notebook, etc.

For some parents (P1, P2, P3), talking to other parents was an informative and supporting activity. Sometimes, they even reported preferring this to talking to a member of the healthcare team. Therefore, some parents requested to modify the activity schedule for their biannual appointments by adding more time to talk with other parents.

For the majority of parents (P1, P2, P3, P4, P5, P9, P10), although physical activity could be an area of concern with their child, other important worries and anxieties concerned infusions, especially in the immediate period after diagnosis. This theme came back often in their discourse. Parents seemed to be lacking information on this topic, especially on an emotional level, e.g., struggling with the technique, inflicting pain to their child during the procedure, ...

Some parents (P7, P10) also mentioned that dealing with the perceptions of others and misconceptions about the illness in the population were a challenge.

Discussion générale

Résumé et synthèse des articles

Dans ce projet, nous avons d'abord traduit le VERITAS-Pro et VERITAS-PRN en français, deux instruments d'importance pour l'étude de l'adhésion au traitement en hémophilie, de ses déterminants et de ses répercussions (Duncan, Kronenberger, Roberson, & Shapiro, 2010). Nous nous sommes assurés que la traduction de cet instrument le rendait accessible autant à une population pédiatrique qu'aux parents et aux professionnels de la santé. Les données psychométriques préliminaires décrites dans notre étude appuient son utilisation. Nous avions également le souci que cette première étude puisse décrire de manière détaillée l'adhésion au traitement auprès de nos participants, à la fois en ce qui a trait aux différents domaines d'adhésion décrits dans les questionnaires, mais également en ce qui concerne les différences observées entre les répondants.

L'adhésion était la plus basse en ce qui concerne la propension des patients et de leurs parents à communiquer avec le centre de traitement lorsqu'ils ont des questions ou des préoccupations. Il est donc essentiel de continuer les efforts pour favoriser une relation de confiance entre patients, parents et soignants. Cependant, il faut aussi considérer que les patients utilisent souvent d'autres sources d'information pour trouver réponse à leurs questions, comme Internet, leurs proches et les dépliants (Cutilli, 2010). Il s'avère donc aussi important de fournir du matériel d'information de qualité et de diriger les parents et les patients vers des sources fiables d'information. Dans cette première étude, les parents rapportaient de meilleurs scores d'adhésion au traitement que ceux rapportés par les enfants et les adolescents et pour les mêmes conditions. Cela soulève l'importance d'obtenir le point de vue à la fois des parents et des patients eux-mêmes, afin de mieux évaluer l'adhésion et de déterminer où se situent les difficultés. Dans la pratique clinique, la complétion du questionnaire VERITAS-Pro ou VERITAS-PRN pourrait permettre de cibler rapidement les domaines d'adhésion plus problématiques ou ceux où il y a de la dissension entre les réponses du parent et de l'enfant pour ensuite, lors d'entrevues cliniques, les évaluer plus en profondeur. Finalement, nous avons aussi confirmé nos hypothèses s'appuyant sur des études antérieures qui proposent que l'adhésion au traitement par injection diminue lorsque les jeunes deviennent autonomes dans leur administration de traitement (Duncan et al., 2012; Geraghty et al., 2006). C’est en s’appuyant sur ces données que nous avons voulu favoriser le développement d’une forme de motivation plus intrinsèque chez les jeunes par notre projet de livret d’information en dernière partie de la thèse.

Le deuxième article qui a comme fondement la théorie du comportement planifié nous a permis d’étudier les comportements de santé importants en hémophilie (le traitement par injection et l’activité physique) et des facteurs psychosociaux qui sont spécifiquement et directement associés à ces comportements. La seconde étude utilisant cette théorie se centrait donc davantage sur les recommandations en tant que telles, à savoir si celles-ci sont appliquées ou non par les patients et dans quelle mesure. Nous voulions savoir si celles-ci sont perçues de manière positive, négative (attitudes), si elles sont encouragées ou non par l’environnement social de l’enfant (normes sociales) et si les patients se sentent capables de les appliquer (contrôle comportemental).

Nous avons été surpris de constater le haut niveau d’adhésion au traitement par injection dans notre échantillon. En effet, pour l’ensemble des répondants (parents et enfants), le score total moyen au VERITAS-Pro (prophylaxie) était de 38.45(13.71) et de 35.95(10.92) au VERITAS-PRN (traitement sur demande). Considérant que le meilleur score possible à cet instrument de 24 (adhésion parfaite) et que le seuil minimal de non-adhésion proposé par la chercheuse qui a développé le VERITAS-Pro est de 57 (aucun seuil minimal n’est proposé pour le VERITAS-PRN), nous pouvons affirmer que l’adhésion rapportée au traitement par injection s’est avérée très élevée dans l’échantillon (Duncan et al., 2010a).

En ce qui concerne l’activité physique, un constat différent s’est imposé. En effet, nous avons pu établir que les participants pratiquaient des activités physiques déconseillées en moyenne un jour et demi par semaine, ce qui est préoccupant considérant les risques qui y sont associés. Ceci nous a amené à concentrer nos efforts sur la compréhension de l’adhésion aux recommandations d’activité physique dans la suite du programme de recherche. Dans cette deuxième étude, il est aussi ressorti que le comportement passé, l’influence subjective des parents et des amis, ainsi que l’attitude des jeunes envers l’activité physique déconseillée était intimement liée à leur intention de pratiquer ces activités. Plus spécifiquement, nous avons découvert que plus les jeunes croyaient que l’évitement des sports déconseillés pouvait aider à la prévention des saignements, moins ils étaient enclins à pratiquer celles-ci. Toutefois, en ce qui a trait à l’activité physique recommandée, les jeunes ne semblaient pas tous saisir l’aspect bénéfique de celle-ci sur la prévention des saignements. Ses bénéfices ont pourtant été bien démontrés dans la littérature et l'activité physique sécuritaire fait partie intégrante de la stratégie de soin en hémophilie (Buzzard, 1996; Negrier et al., 2013). Cela nous amène à constater qu'il existe des écarts entre l'information qui est connue et véhiculée par les soignants et ce que les jeunes patients peuvent retenir et intégrer comme information et qui guide leur comportement au quotidien. Cet article nous a en somme permis de mieux comprendre la perspective des jeunes sur les recommandations qui leur sont données par leur équipe de soin. Il semble que la pratique d'activité déconseillée serait plus susceptible d'être influencée par des interventions psychosociales étant donné qu'elle était davantage associée aux facteurs psychosociaux étudiés.

La troisième étude, basée sur la théorie du sens commun, a permis d'évaluer les perceptions de la maladie chez les jeunes atteints d'hémophilie et leur relation aux différents profils d'activité physique. La théorie du sens commun est un modèle dynamique qui s'intéresse davantage au rapport subjectif qu'entretient le patient avec sa maladie (Leventhal & Ian, 2012). Ainsi, plutôt que de s'intéresser à un comportement en particulier et à ses prédicteurs proximaux, la théorie élargit notre compréhension en nous amenant à considérer le modèle interne, ou la façon dont la personne conceptualise sa maladie, à savoir si celle-ci s'intègre aisément à sa vie ou au contraire est perçue comme une menace aux buts importants pour la personne (Cameron & Leventhal, 2003). Le modèle nous amène à nous questionner sur les stratégies d'autorégulation que les patients mettent en place pour faire face à la menace, à la fois sur le plan comportemental (actions), mais également sur le plan affectif (stratégies de régulation émotionnelle).

Ainsi, l'article 3 nous a permis de nous pencher davantage sur les composantes émotionnelles reliées à l'adhésion aux recommandations sur l'activité physique. Cet aspect est peu élaboré dans la théorie du comportement planifié, utilisée dans la deuxième étude, si ce n'est que l'attitude comporte une composante émotionnelle (Buzzard, 1996). Nous avons été surpris de constater dans cette troisième étude que plusieurs jeunes, malgré une grande confiance envers l'efficacité de leur traitement et en leur capacité à contrôler leur condition de santé, pouvaient être grandement affectés par l'hémophilie sur le plan émotionnel. La qualité de vie des patients s'est beaucoup améliorée depuis les années 1980 avec la possibilité d'administrer les injections à la maison (Teitel et al., 2004). Notre étude soulève un point important, soit que l'amélioration des traitements disponibles n'est pas nécessairement synonyme de bien-être psychologique. En d'autres mots, ces avancées médicales ne doivent pas faire en sorte d'abandonner les efforts pour mieux détecter la détresse chez certains patients ou de les prémunir de recevoir l'aide psychosociale lorsque cela est approprié, voire nécessaire. En effet, dans une étude américaine sur 200 patients adultes atteints d'hémophilie, une grande proportion de ceux-ci (28%) présentait des niveaux modérés à sévères de dépression (Witkop et al., 2019). Une autre étude américaine toujours chez les adultes atteints d'hémophilie a aussi démontré de hauts niveaux de dépression chez cette population (37% remplissaient les critères) (Iannone et al., 2012). Chez les adultes atteints de cette condition, les niveaux de dépression étaient aussi associés à une moins bonne adhésion au traitement et à un plus grand nombre de saignements dans la dernière année (Pinto et al., 2018; Witkop et al., 2019). Une étude chez les enfants atteints d'hémophilie montre quant à elle des résultats allant dans le même sens que les nôtres, soit de plus hauts niveaux de symptômes dépressifs chez ceux-ci comparativement aux enfants en santé, ainsi que d'autres indices de santé mentale plus faibles (Trzepacz, Vannatta, Davies, Stehbens, & Noll, 2003).

Sur le plan comportemental, dans notre étude, et tel que nous l'avions prédit, le groupe des jeunes ayant un profil d'adhésion plus faible avait aussi des perceptions de la maladie plus négatives, percevait plus de symptômes et était davantage préoccupé par la maladie que les jeunes qui adhéraient aux recommandations. Ces résultats sont particulièrement intéressants puisque les professionnels de la santé et les proches des patients pourraient penser que les jeunes prennent la décision de pratiquer une activité physique dangereuse parce qu'ils sont peu inquiets par rapport à leur condition, et donc, peu mobilisés dans leur traitement. Une telle hypothèse pourrait inciter les proches et les professionnels à mettre l'emphase sur les risques associés à certains comportements dans leur communication avec les patients. Or, on doit user de prudence dans l'utilisation d'un discours visant à éveiller davantage de peur chez les patients. Une méta-analyse sur les messages de peur, axés sur les conséquences négatives d'un comportement, dans le domaine de la santé, conclut que ceux-ci ne sont pas toujours appropriés (Witte & Allen, 2000). En effet, lorsque les messages de peur ne réussissent pas à convaincre le patient que les mesures à prendre pour éviter des conséquences négatives sont efficaces ou lorsque le patient se sent incapable de mettre en place les actions nécessaires, le message est plus susceptible de générer des réponses défensives qui interfèrent avec les actions comme du déni ou une réaction du type « ils essaient juste de me manipuler, je vais les ignorer » (Witte & Allen, 2000). Ainsi, nous pouvons nous questionner à savoir si, selon la perception des jeunes pratiquant déjà des activités risquées, il est faisable et réaliste d'arrêter complètement et de changer rapidement cette habitude. En lien avec nos résultats, il est probablement très important de créer une ouverture dans les échanges avec les jeunes pour qu'ils se sentent à l'aise de partager leurs émotions, leurs perceptions et leurs dilemmes intérieurs plutôt que d'amener de l'information trop rapidement ou d'axer simplement sur les conséquences négatives que peuvent avoir leur comportement. Accorder une plus grande place aux perceptions, aux buts, aux valeurs et aux émotions des patients fait d'ailleurs partie des dimensions importantes d'une approche davantage centrée sur le patient, approche qui est encouragée par les organisations en médecine depuis plusieurs années (Epstein, Fiscella, Lesser, & Stange, 2010; Tzelepis, Sanson-Fisher, Zucca, & Fradgley, 2015).

Dans la suite du programme de thèse, nous avons utilisé les concepts du soutien à l'autonomie afin d'adresser les enjeux de motivation et d'adhésions chez les patients (Cameron & Leventhal, 2003). Ceci reflète le souci que nous avions de rendre ce projet de recherche utile non seulement en ce qui concerne la recherche empirique, mais également sur le plan clinique. Plutôt que d'intervenir directement avec les enfants dans une intervention en milieu hospitalier, nous avons fait le choix d'enseigner ces principes aux parents sous forme de livret d'information. Ainsi, nous voulions que l'intervention se produise au travers des interactions parent-enfant quotidiennes et dans une perspective à long terme. Ce type d'intervention permet aussi de répondre aux besoins de soutien que peuvent ressentir les parents confrontés aux décisions difficiles de la vie de tous les jours en lien avec l'hémophilie de leur enfant. En effet, les parents d'enfants atteints de maladie chronique font face à plusieurs dilemmes importants : protéger leur enfant versus favoriser encourager son développement et son autonomie, partager l'information médicale à leur enfant versus le protéger d'émotions désagréables, et récompenser l'implication de leur enfant versus la mise en place de limites importantes (Seo et al., 2019). Dans le discours des parents d'enfants atteints de maladie chronique, on comprend que les parents doivent eux-mêmes traverser un processus de transition dans lequel ils doivent apprendre à transférer graduellement certaines tâches et accepter que leur enfant acquière graduellement plus d'autonomie, et ce, même si cela implique que celui-ci puisse parfois faire des erreurs (Heath, Farre, & Shaw, 2017). Ainsi, les parents doivent apprendre à tolérer leurs inquiétudes. Aider les parents à mieux comprendre l'importance de soutenir l'autonomie de leur enfant et les façons pour y arriver est nécessaire et c'est cette prémisse qui a guidé l'écriture des articles 4 et 5.

Pour faire le pont entre les différentes théories utilisées dans la présente thèse, les concepts du soutien à l'autonomie de la théorie de l'autodétermination peuvent également s'arrimer aux modèles théoriques vus précédemment. En effet, en ce qui a trait à la théorie du comportement planifié, le modèle du soutien à l'autonomie stipule qu'il ne faut pas seulement viser à augmenter l'intention. En effet, à la lumière des écrits sur le soutien à l'autonomie, nous comprenons qu'il y aurait différents types d'intention (de motivation), certaines formes plus intégrées que d'autres et qui sont plus susceptibles de se maintenir dans le temps (Deci & Ryan, 2017). De la même façon, il faut porter un regard attentif sur les différentes formes que peuvent prendre l'attitude, les normes subjectives et le contrôle comportemental. Par exemple, d'autres auteurs proposent d'utiliser le concept de soutien à l'autonomie pour comprendre comment et dans quelle mesure les normes subjectives peuvent influencer le comportement. En effet, le soutien à l'autonomie est une forme de soutien efficace et non pressurisante, par opposition à d'autres formes de pressions sociales qui seraient moins adaptées, comme les récompenses et les punitions (Chatzisarantis et al., 2008). Bien que la littérature sur les liens entre la théorie du comportement planifié et le soutien à l'autonomie soit très mince, ces modèles ont été déjà utilisés en complémentarité. Par exemple, une étude a démontré que des stratégies de soutien à l'autonomie peuvent avoir un impact sur l'intention de pratique de l'activité physique et sur l'attitude telle que décrite dans la théorie du comportement planifié (Hagger, Chatzisarantis, Barkoukis, Wang, & Baranowski, 2005).

Plus concrètement, le livret d'information que nous avons développé encourage les parents à s'intéresser au point de vue de leur enfant, sans essayer de modifier ce que les enfants pensent et ressentent face à la maladie. Ils sont encouragés à agir comme support émotionnel pour leurs enfants. Nous émettons l'hypothèse qu'une telle approche puisse favoriser une attitude plus positive des enfants à l'égard des restrictions, de manière à ce qu'il se sentent compris, validés, et qu'ils aient l'espace pour trouver leurs propres solutions. Il est également possible qu'une telle approche ait un impact sur les normes subjectives en amenant les parents à être plus à l'aise dans leur capacité à interdire certaines activités physiques à leurs enfants. Le soutien à l'autonomie permet également aux parents d'apprendre à mieux tolérer et contenir les réactions émotionnelles de leurs enfants et donc, à maintenir les règles qui ont été établies en lien avec l'activité physique, même lorsque l'enfant réagit moins bien initialement (tristesse, déception, colère, etc.). Nous pourrions aussi émettre l'hypothèse que l'implication graduelle de l'enfant dès son jeune âge dans les différents aspects du plan de traitement est susceptible de favoriser une meilleure perception de contrôle comportemental au fil du temps.

La théorie de l'autodétermination peut aussi être considérée comme en phase avec la théorie du sens commun, bien que les liens qui les unissent soient moins étudiés dans la littérature. En effet, le développement d'une meilleure régulation émotionnelle a été abordé dans les deux théories (Cameron & Leventhal, 2003; Deci & Ryan, 2004). Le modèle du soutien à l'autonomie encourage les parents à communiquer avec leur enfant de manière à ce que ces derniers se sentent écoutés et soutenus et qu'ils aient l'espace pour vivre leurs émotions et apprendre à les réguler. Par exemple, on encourage les parents à ne pas apporter de solutions trop rapidement, à écouter, à tolérer le vécu émotionnel de leur enfant et à l'aider à mettre des mots sur celui-ci. Le soutien à l'autonomie des parents favoriserait d'ailleurs un attachement sécure chez l'enfant (Whipple, Bernier, & Mageau, 2011). Le modèle de l'autodétermination invite également l'enfant à être créatif et à trouver ses propres solutions lorsqu'il rencontre un problème ou un dilemme, et donc, nous pouvons penser qu'il puisse l'aider à développer un plan d'action efficace et personnel, tel qu'encouragé par la théorie de l'autodétermination. Ainsi, une approche axée sur le soutien à l'autonomie pourrait potentiellement avoir un effet sur les deux voies de régulation explicitées dans le modèle du sens commun, soit la voie comportementale et la voie émotionnelle (Cameron & Leventhal, 2003). En somme, nous pouvons considérer que le livret que nous avons développé invite les parents à s'intéresser aux perceptions de la maladie de leur enfant en les questionnant davantage, en les amenant à s'exprimer à propos de la maladie. Il serait alors possible pour les parents de découvrir si leur enfant entretient des perceptions de la maladie moins adaptées. Le livret pourrait les aider à trouver des façons d'aider leur enfant à mieux réfléchir au sujet de la maladie, sans imposer leurs propres idées et en soutenant leur processus d'autorégulation émotionnelle. Ce livret pourrait être particulièrement pertinent pour les parents des jeunes patients qui ne respectent pas les recommandations autour de l'activité physique, puisqu'il est ressorti de notre troisième étude que ces jeunes voyaient plus négativement l'hémophilie et vivaient davantage de préoccupations.

Contribution scientifique et clinique

En somme, ce programme de thèse nous a permis d'éclaircir où se situent les difficultés d'adhésion chez les jeunes atteints d'hémophilie. En réunissant l'apport de différentes théories à la compréhension du phénomène de l'adhésion aux recommandations, nous avons fait ressortir des facteurs qui sont susceptibles de contribuer de manière positive et négative à l'adhésion des jeunes atteints d'hémophilie sévère. De plus, ce programme de thèse permet de soulever différents enjeux psychosociaux à adresser cliniquement, notamment l'impact émotionnel que la maladie peut avoir sur les patients, en dépit du fait qu'un traitement efficace soit disponible pour les jeunes patients canadiens. À ce jour, peu d'études psychosociales sur l'adhésion aux recommandations en hémophilie existent. Celles qui ont été menées se sont surtout centrées sur l'adhésion au traitement par injection en utilisant le plus souvent des sondages et des études qualitatives plutôt que des études corrélationnelles. D'ailleurs, l'instrument de référence pour mesurer l'adhésion au traitement de manière efficace, le VERITAS, n'était pas encore disponible en français. Ce projet de recherche est aussi novateur par son emphase sur l'adhésion aux recommandations d'activité physique, emphase quasi absente des travaux de recherche antérieurs. Pourtant, les limitations entourant l'activité physique ressortent souvent comme étant particulièrement problématiques et difficiles pour les enfants et les adolescents dans les études qualitatives (Flaherty, Schoeppe, Kruse-Jarres, & Konkle, 2018; Nazzaro et al., 2006; Williams & Chapman, 2011). Nous avons aussi été en mesure de constater les besoins en termes d'intervention dans cette population, non seulement celles visant à améliorer l'adhésion aux recommandations entourant l'activité physique, mais également celles visant à favoriser le bien-être émotionnel de plusieurs jeunes patients.

Parallèlement à la contribution scientifique de cette thèse, le livret d'information que nous avons développé en collaboration avec l'équipe de soin et les parents, en s'appuyant sur les connaissances que nous avons pu acquérir tout au long de ce projet de recherche, sera disponible au CHU Sainte-Justine. Il est impératif que la recherche scientifique en psychologie de la santé mène à amélioration des soins et permette d'apporter un soutien concret aux patients et à leurs proches. Il y a souvent un écart important entre les avancées scientifiques et les avancées cliniques. Nous sommes conscients qu'un simple livret d'information n'est pas à la hauteur des besoins de ces patients, mais nous espérons qu'il puisse contribuer de manière significative à améliorer leur bien-être et leur qualité de vie. Il est aussi important de souligner que celui-ci vient accompagner les activités et les interventions de qualité données les professionnels de la santé, qui sont déjà disponibles au CHU Sainte-Justine. Une revue de la littérature sur les interventions psychosociales empiriquement validées en hémophilie met en lumière le peu d'études disponible dans ce domaine (Palareti, Melotti, Cassis, Nevitt, & Iorio, 2020). Cela fait contraste à la riche littérature sur les interventions psychosociales dans d'autres problématiques de santé chez les enfants tel que le cancer (Waldron, Janke, Bechtel, Ramirez, & Cohen, 2013; Walker, Martins, Aldiss, Gibson, & Taylor, 2016). Un apport de nos travaux concerne aussi le fait que les concepts du soutien à l'autonomie ont souvent été étudiés dans le cadre de la relation patient-soignant, sans nécessairement être étudiés dans le cadre relation parent-patient atteint d'une maladie chronique (Vasquez, Patall, Fong, Corrigan, & Pine, 2016; Williams & Deci, 2001b; Williams, McGregor, King, Nelson, & Glasgow, 2005). Nous espérons inciter d'autres chercheurs à reprendre les concepts du soutien à l'autonomie pour travailler d'autres aspects de la relation parent-enfant en psychologie de la santé. Quant au livret d'information que nous avons développé, il serait intéressant que les éléments de contenu puissent être repris pour développer des contenus audiovisuels, comme dans les messages diffusés sur écran dans les salles d'attente, car ceux-ci ont déjà été démontrés comme étant efficaces pour améliorer les connaissances et sont généralement appréciés des usagers (Cockington, 1995; Gignon, Idris, Manaouil, & Ganry, 2012).

Finalement, nous avons pu effleurer en discussion de cette thèse l'intégration des différents modèles théoriques. Nous espérons que la façon avec laquelle nous avons utilisé les concepts du soutien à l'autonomie pour pallier certaines lacunes concernant le manque de directives sur l'intervention dans les théories sociocognitives utilisées suscitera l'intérêt des chercheurs et encouragera la communauté scientifique à viser une meilleure intégration de ces différentes traditions théoriques trop souvent utilisées de manière hermétique dans la littérature.

Limites et perspectives futures

Il faut reconnaître les limites du programme de la thèse, surtout en ce qui a trait aux limitations dans la généralisation des résultats. En effet, les méthodes corrélationnelles et les questionnaires autorapportés ne permettent d'établir un lien de causalité et sont soumises à l'influence de la désirabilité sociale et à des biais cognitifs. La théorie du sens commun étant un modèle dynamique et bidirectionnel, elle peut aussi amener de la confusion quant à la direction des liens de cause à effet. Nous nous sommes assurés que nos articles reflètent bien les limites de la méthodologie utilisée.

Nous avons également été limités dans nos analyses par la taille de l'échantillon de participants. Cependant, considérant que l'hémophilie est une maladie rare, le recrutement d'échantillons importants dans ce domaine est très difficile. En termes de statistiques, en 2018, il y avait seulement 423 patients canadiens de moins de 18 ans atteints d'hémophilie sévère A ou B (Canadian Hemophilia Registry, 2018). Pour les articles 1 à 3, nous avons contacté l'ensemble des participants qui satisfaisaient les critères d'inclusion et d'exclusion à l'hôpital Sainte-Justine. Le taux de participation pour les diverses études était excellent, ce qui démontre la motivation des patients et de leurs proches à faire avancer la recherche psychosociale en hémophilie.

Sur le plan théorique, chacune des théories utilisées comporte en elle-même des limitations qui ont été l'objet de critiques pertinentes au fil du temps. Par exemple, selon certaines critiques, les théories sociocognitives comme la théorie du comportement planifié et la théorie du sens commun assumeraient à tort que les individus font des choix rationnels. Fishbein et Ajzen ont fourni une réponse pertinente à cette critique en expliquant que le modèle n'assume pas que les choix sont rationnels, mais que les comportements des gens suivent une logique interne consistante, même si celle-ci peut être basée sur des croyances irrationnelles (Fishbein & Ajzen, 2011). Il n'a pas été possible pour nous d'évaluer en détail, dans l'article 2, ces croyances spécifiques (facteurs distaux) qui peuvent contribuer à l'attitude, au sentiment de contrôle et à la norme subjective (facteurs les plus proximaux du comportement). Par exemple, des croyances peuvent être de type « les saignements ne sont pas si graves que les adultes le laissent croire » ou « je n'aurai plus d'amis si je ne pratique pas le football ». Pour évaluer ces croyances, il aurait fallu proposer un large éventail de questions, ce qui aurait demandé un échantillon de patients substantiellement plus grand afin de s'assurer d'une puissance statistique suffisante.

Notre façon d'obtenir davantage de spécificité sur les perceptions et les croyances a été d'utiliser une seconde théorie, le modèle du sens commun de la théorie de l'autorégulation (article 3), qui est un modèle plus dynamique et plus vaste que la théorie du comportement planifié, et apporte peut-être une meilleure compréhension de la subjectivité des patients face à leur maladie. Cependant, ce modèle a souvent été critiqué comme ayant une piètre validité prédictive (Aujla et al., 2016). De notre côté, cette complexité a généré des questionnements qui pourraient être investigués dans des recherches futures. Pour ne nommer que cela, l'article 2 nous amène à nous questionner sur ce qui explique que les individus non adhérents aux recommandations d'activité physique soient plus préoccupés par leur maladie que ceux qui sont plus adhérents. Il serait aussi intéressant de comprendre la nature des préoccupations qui habitent les patients en lien avec leur maladie. Pour contrer les difficultés reliées au petit échantillonnage, ces questions pourraient être explorées sous un angle qualitatif. L'utilisation des nouvelles technologies, comme les études en ligne, et la concertation entre les différents chercheurs dans le domaine permettraient d'obtenir des échantillons substantiellement plus grands.

Il est aussi à noter que nous avons fait le choix de nous concentrer sur les facteurs individuels reliés à l'adhésion. Notre souci était de comprendre comment les jeunes se positionnent face à leur condition et aux recommandations de traitement. Nous sommes conscients que les parents occupent une place très importante dans le processus d’autonomisation de leurs enfants et d’apprentissage de l’autosoin, et que l’étude des facteurs parentaux reliés à l’adhésion au traitement ou à l’adaptation des jeunes à leur maladie est tout à fait pertinente.

Nous tenons aussi à faire mention que nos données ne sont valables que dans le contexte de pays développés. En effet, dans les pays en voie de développement, malheureusement, l’accessibilité à la prophylaxie est très limitée et les patients souffrent de graves complications de l’hémophilie. À titre de statistique, 36 % des patients (ceux qui vivent dans les pays les plus industrialisés) consomment 96 % des facteurs VIII disponibles et la moitié des enfants hémophiles dans les pays en voie de développement décèdent avant l’âge de 10 ans (Fédération mondiale de l'hémophilie, 2020). À cet effet, nous désirons faire connaître le travail de la Fédération mondiale de l’hémophilie qui, grâce à des levées de fond, permet à des milliers d’enfants de recevoir un traitement approprié.

Conclusion

En conclusion, ce programme de thèse visait à combler certaines lacunes dans la littérature actuelle en hémophilie, en particulier en ce qui a trait aux enjeux psychosociaux susceptibles d’avoir un impact sur les comportements et sur la santé des jeunes atteints de cette maladie. Cette thèse nous a permis à la fois de dégager des prédicteurs potentiels de l’adhésion au traitement par injection et de la pratique d’activité physique. Nous avons pu comprendre que les difficultés d’adhésion se situaient davantage sur le plan de l’activité physique. L’exploration du point de vue des jeunes patients sur leur condition de santé et sur les différentes mesures qui visent à diminuer le nombre de saignements auxquels ils auront à faire face nous a permis de mieux comprendre leurs besoins et de développer des stratégies d’intervention pour favoriser une meilleure adaptation à la maladie chez ces patients, mais également chez leurs parents qui sont responsables d’une bonne partie de la gestion de la maladie et de son traitement. Peu d’outils sont actuellement disponibles pour guider les parents d’enfants atteints d’hémophilie sur les façons de communiquer efficacement au sujet des différents enjeux de la maladie. Nous espérons que nos travaux pourront susciter chez d’autres chercheurs un intérêt pour ces enjeux en hémophilie ou à adapter les idées d’intervention que nous avons développées à d’autres conditions de santé. Entre autres, les recherches futures pourraient s’attarder aux perceptions de la maladie sur de plus vastes échantillons afin de confirmer les résultats que nous avons obtenus, soit que les patients dont le profil d’adhésion était plus fragile entretenaient également des perceptions plus négatives au sujet de la maladie et vivaient davantage de préoccupations en lien avec la maladie. Les recherches pourraient également tenter d’identifier les croyances sous-jacentes à ces perceptions plus négatives. En ce qui a trait à l’intervention, il serait intéressant de vérifier les effets du livret d’information sur la motivation des jeunes à suivre les recommandations au sujet de l’activité physique et sur l’adhésion à celles-ci. De plus, d’autres formats d’interventions visant à augmenter les compétences parentales de soutien à l’autonomie pourraient être explorés et évalués afin de voir si ceux-ci peuvent contribuer à une meilleure adaptation à la maladie et aux recommandations médicales qui y sont associées.

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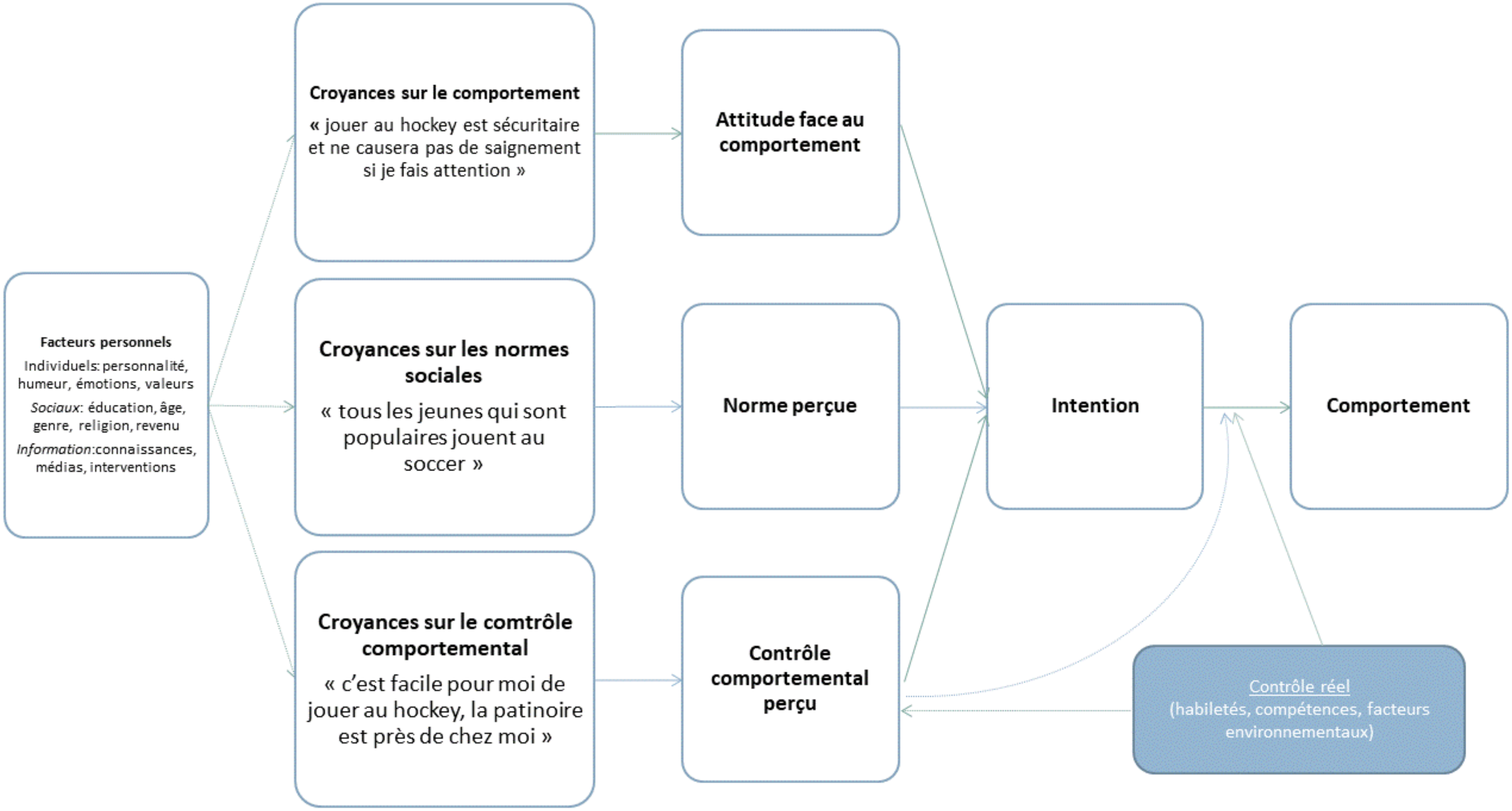
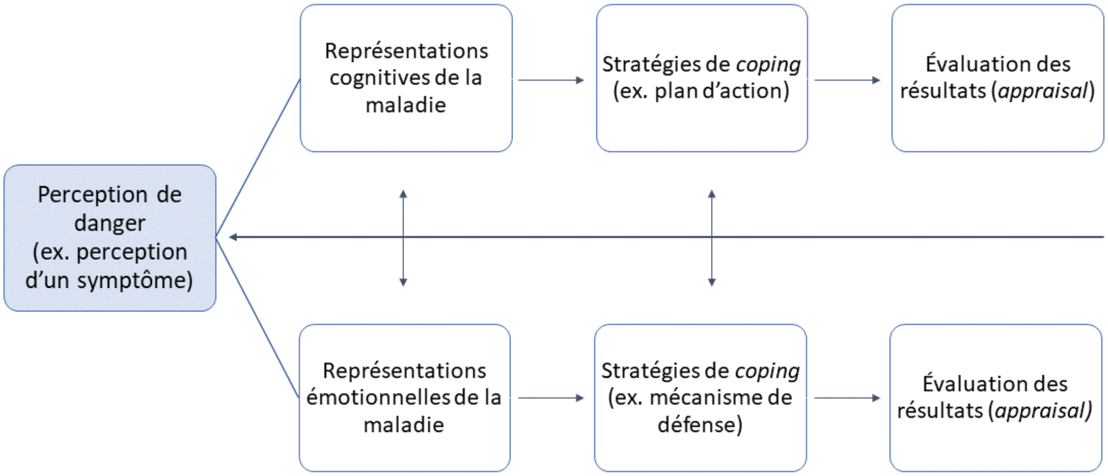
Annexe A

Figure 2. Théorie du comportement planifié (adapté de Fishbein & Ajzen, 2011)



Annexe B

Figure 1. Modèle du sens commun de la théorie de l'autorégulation (adapté de Cameron & Leventhal, 2003)



Annexe C

Attitudes et croyances concernant les comportements d'autosoin
Questionnaire pour les jeunes

I. ACTIVITÉ PHYSIQUE, EXERCICE ET SPORT

Les questions suivantes portent sur l'exercice, l'activité physique et le sport.

Croyances comportementales, attentes concernant les résultats, coûts et bénéfices, conséquences perçues

1. Tu vois le fait de pratiquer régulièrement une activité physique **recommandée** de manière…

négative **1 2 3 4 5 6 7** *positive*

2. Tu vois le fait de pratiquer régulièrement une activité physique **déconseillée** de manière…

négative **1 2 3 4 5 6 7** *positive*

3. Pratiquer régulièrement une activité physique **recommandée** va prévenir les saignements dans tes articulations (coudes, genoux, chevilles).

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

4. Pratiquer régulièrement une activité physique **déconseillée** va augmenter les saignements dans tes articulations (coudes, genoux, chevilles).

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

5. Prévenir les saignements dans tes articulations (coudes, genoux, chevilles) est...

pas important **1 2 3 4 5 6 7** *important*

6. Tes parents apprécient ou approuvent que tu pratiques une activité physique **recommandée**.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

7. Tes parents apprécient ou approuvent que tu pratiques une activité physique **déconseillée**.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

8. En ce qui concerne l'activité physique, tu veux écouter tes parents (faire ce qu'ils veulent que tu fasses).

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

9. Tes amis apprécient que tu pratiques une activité physique **recommandée**.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

10. Tes amis apprécient que tu pratiques une activité physique **déconseillée.**

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

Sentiment d'auto-efficacité

11. À quel point es-tu confiant(e) (ou te sens-tu capable) de pouvoir pratiquer régulièrement une activité physique **recommandée**?

pas du tout confiant(e) **1 2 3 4 5 6 7** *tout à fait confiant(e)*

12. À quel point es-tu confiant(e) (ou te sens-tu capable) de pouvoir éviter de pratiquer régulièrement une activité **déconseillée**?

pas du tout confiant(e) **1 2 3 4 5 6 7** *tout à fait confiant(e)*

Intentions d'accomplir le comportement

13. Dans l'avenir, tu as l'intention de pratiquer une activité physique **recommandée.**

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

14. Dans l'avenir, tu as l'intention de pratiquer une activité physique **déconseillée**.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

II. GESTION DE L'INJECTION DE CONCENTRÉ DE FACTEUR

Important : les questions suivantes portent sur la prophylaxie, donc sur les injections faites sur un horaire régulier et ne concernent **pas** le traitement sur demande.

Auto-administration des injections préventives

15. Est-ce qu'il t'arrive **d'autoadministrer (faire toi-même)** ton injection préventive de concentré de facteur (prophylaxie)?

a) Oui ☐
b) Non ☐ Si non, passe à la question 60.

16. Tu **autoadministres (fais toi-même)** ton injection préventive de concentré de facteur (prophylaxie) …

a) Toujours ☐
b) Parfois ☐
c) Jamais ☐

17. Es-tu supervisé(e) par un adulte lorsque tu **autoadministres (fais toi-même)** ton injection préventive de concentré de facteur (prophylaxie)?

a) Toujours ☐
b) Parfois ☐
c) Jamais ☐

Croyances comportementales, attentes concernant les résultats, coûts et bénéfices, conséquences perçues

18. Tu vois le fait de toujours recevoir (ou autoadministrer) tes injections préventives (prophylaxie) tel que recommandé de manière…

négative **1 2 3 4 5 6 7** *positive*

19. Toujours recevoir (ou autoadministrer) tes injections préventives (prophylaxie) tel que recommandé est bon pour ta santé.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

20. Toujours recevoir (ou autoadministrer) tes injections préventives (prophylaxie) tel que recommandé va prévenir les saignements dans tes articulations (coudes, genoux, chevilles).

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

Pressions normatives perçues

21. Tes **parents** apprécient ou approuvent que tu reçoives (ou autoadministres) toujours tes injections préventives (prophylaxie) tel que recommandé.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

22. En ce qui concerne tes injections préventives (prophylaxie), tu veux écouter tes **parents** (faire ce qu'ils veulent que tu fasses).

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

23. Tes **amis** apprécient ou approuvent que tu reçoives (ou autoadministré) tes injections préventives (prophylaxie) tel que recommandé.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

☐ Mes amis ne savent pas que je reçois des injections

Sentiment d'auto-efficacité

24. À quel point es-tu confiant(e) ou te sens-tu capable de toujours pouvoir recevoir (ou autoadministrer) tes injections préventives (prophylaxie) tel que recommandé?

pas du tout confiant(e) **1 2 3 4 5 6 7** *tout à fait confiant(e)*

Intentions d'accomplir le comportement

25. Dans l'avenir, tu as l'intention de recevoir (ou autoadministrer) tes injections préventives (prophylaxie) tel que recommandé.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

Important : les questions suivantes portent sur le traitement sur demande, donc sur les injections qui sont faites de façon ponctuelle et ne concernent **pas** la prophylaxie.

26. Est-ce qu'il t'arrive **d'autoadministré (faire toi-même)** ton injection « sur demande » de concentré de facteur **(après une chute ou une blessure, ou lorsque tu remarques que tu as de la douleur, de la difficulté à marcher ou à utiliser un membre)** ?

a) Oui ☐
b) Non ☐ Si non, passe à la question 89.

27. Tu **autoadministres (fais toi-même)** ton injection « sur demande » de concentré de facteur **(après une chute ou une blessure, ou lorsque tu remarques que tu as de la douleur, de la difficulté à marcher ou à utiliser un membre)** …

a) Toujours ☐
b) Parfois ☐
c) Jamais ☐

28. Tu es supervisé(e) par un adulte lorsque tu **autoadministres (fais toi-même)** ton injection « sur demande » de concentré de facteur **(après une chute ou une blessure, ou lorsque tu remarques que tu as de la douleur, de la difficulté à marcher ou à utiliser un membre)**…

a) Toujours ☐
b) Parfois ☐
c) Jamais ☐

29. Toujours recevoir (ou autoadministrer) tes injections « sur demande » lorsque tu en as besoin **(après une chute ou une blessure, ou lorsque tu remarques que tu as de la douleur, de la difficulté à marcher ou à utiliser un membre)** va prévenir les saignements dans tes articulations (coudes, genoux, chevilles).

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

Pressions normatives perçues

30. **Tes parents** pensent que tu devrais toujours recevoir (ou autoadministrer) tes injections « sur demande » tel que recommandé **(après une chute ou une blessure, ou lorsque tu remarques que tu as de la douleur, de la difficulté à marcher ou à utiliser un membre)**.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

31. En ce qui concerne tes injections « sur demande », tu veux écouter **tes parents** (faire ce qu'ils veulent que tu fasses).

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

32. **Tes amis** pensent que tu devrais toujours recevoir (ou autoadministrer) tes injections « sur demande » tel que recommandé **(après une chute ou une blessure, ou lorsque tu remarques que tu as de la douleur, de la difficulté à marcher ou à utiliser un membre)**.

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

☐ Mes amis ne savent pas que je reçois des injections

33. En ce qui concerne tes injections précoces, tu veux écouter **tes amis** (faire ce qu'ils veulent que tu fasses).

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

☐ Mes amis ne savent pas que je reçois des injections

Sentiment d'auto-efficacité

34. À quel point es-tu confiant(e) ou te sens-tu capable de toujours pouvoir recevoir (ou autoadministrer) tes injections « sur demande » toutes les fois où tu en en besoin **(après une chute ou une blessure, ou lorsque tu remarques que tu as de la douleur, de la difficulté à marcher ou à utiliser un membre)**?

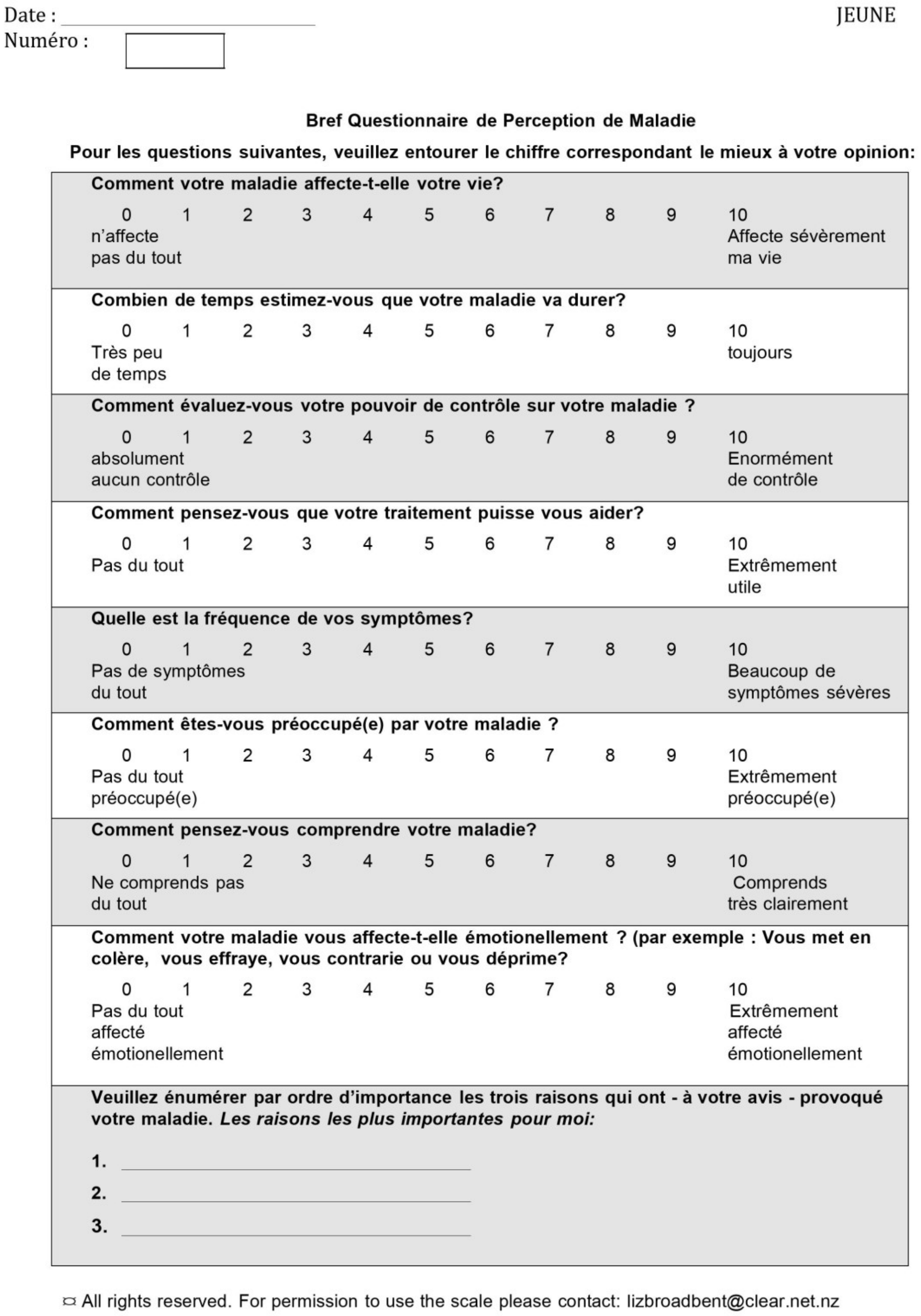
pas du tout confiant(e) **1 2 3 4 5 6 7** *tout à fait confiant(e)*

Intentions d'accomplir le comportement

35. Dans l'avenir, tu as l'intention de recevoir (ou autoadministrer) tes injections « sur demande » chaque fois où tu en as besoin **(après une chute ou une blessure, ou lorsque tu remarques que tu as de la douleur, de la difficulté à marcher ou à utiliser un membre).**

pas du tout d'accord **1 2 3 4 5 6 7** *tout à fait d'accord*

Annexe D



Date : ____________________ JEUNE

Numéro :

Bref Questionnaire de Perception de Maladie

Pour les questions suivantes, veuillez entourer le chiffre correspondant le mieux à votre opinion:

Comment votre maladie affecte-t-elle votre vie?

0	1	2	3	4	5	6	7	8	9	10
n'affecte pas du tout										Affecte sévèrement ma vie

Combien de temps estimez-vous que votre maladie va durer?

0	1	2	3	4	5	6	7	8	9	10
Très peu de temps										toujours

Comment évaluez-vous votre pouvoir de contrôle sur votre maladie ?

0	1	2	3	4	5	6	7	8	9	10
absolument aucun contrôle										Enormément de contrôle

Comment pensez-vous que votre traitement puisse vous aider?

0	1	2	3	4	5	6	7	8	9	10
Pas du tout										Extrêmement utile

Quelle est la fréquence de vos symptômes?

0	1	2	3	4	5	6	7	8	9	10
Pas de symptômes du tout										Beaucoup de symptômes sévères

Comment êtes-vous préoccupé(e) par votre maladie ?

0	1	2	3	4	5	6	7	8	9	10
Pas du tout préoccupé(e)										Extrêmement préoccupé(e)

Comment pensez-vous comprendre votre maladie?

0	1	2	3	4	5	6	7	8	9	10
Ne comprends pas du tout										Comprends très clairement

Comment votre maladie vous affecte-t-elle émotionellement ? (par exemple : Vous met en colère, vous effraye, vous contrarie ou vous déprime?

0	1	2	3	4	5	6	7	8	9	10
Pas du tout affecté émotionellement										Extrêmement affecté émotionellement

Veuillez énumérer par ordre d'importance les trois raisons qui ont - à votre avis - provoqué votre maladie. ***Les raisons les plus importantes pour moi:***

1. ____________________

2. ____________________

3. ____________________

Adapté de Broadbent (2010)