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Leg pains in CHD: a distressing symptom of a wider problem

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Abstract

Purpose: Anecdotal reports suggest that children and young adults with CHD frequently experience pain in their legs. The purpose of this pilot study, performed by Little Hearts Matter patient organisation, was to assess the burden of leg pains in this group and begin to investigate associated factors and consequences for daily living. *Methods:* An internet-based survey was distributed by Little Hearts Matter patient organisation. After anonymisation and collation, responses were analysed and compared with their healthy siblings. *Results:* Of the 220 patients who responded, 94% reported leg pains compared with 30% of siblings ($n = 107$; $p < 0.001$). In respondents, pain was typically reported to occur in the lower legs or around the knees or ankles, often associated with crying and screaming (49.0%) and most commonly occurring at night-time (82.0%). Individuals taking aspirin and those who were more active were more likely to report leg pains. Older age was associated with leg pain that occurred with stress ($p = 0.02$) and at night ($p = 0.05$). Analgesia (64.1%) or massage (53.9%) was the preferred option for alleviation. There was no gender bias, association with diagnosis, surgical history, and/ or relationship with diagnosed orthopaedic issues. *Conclusion:* Leg pains are more frequent in those with CHD compared with their healthy siblings. Aetiology is uncertain, but pains share many common characteristics with benign "growing pains".

Mortality in childhood from CHD has reduced markedly over recent decades owing to advances in surgical and intensive care management. As a result, morbidity metrics are increasingly important in evaluating the outcome of CHD treatment. Concurrently, increasing access to the internet and communication through social media has had an impact on how those affected by CHD seek information and manage their own healthcare. Online discussion among patients has begun to reveal common but under-reported issues that have an impact on daily living and may also indirectly influence healthcare outcomes.[1,2]

Evidence from those living with cystic fibrosis suggests that online support enables young people and parents to share experiences of living with long-term conditions and thus develop expertise to empower them in interactions with healthcare professionals.[3] The understandable caution with which clinicians have approached social media has more recently been rivalled by the recognition of the opportunity to better understand the patient experience and thus focus efforts to improve outcomes.[4,5]

In this study, we responded to anecdotal, but common, complaints of leg pains in children and young adults with CHD by carrying out a questionnaire investigating the key characteristics and circumstances of the pain, as well as methods used for alleviation.

Materials and methods

Study population

Little Hearts Matter is a United Kingdom-registered charity dedicated to supporting and enabling families of children and adults with a diagnosis of a single-ventricle heart conditions (www.lhm.org.uk). Through a shared Facebook site, the charity identified concerns among patients and their carers regarding the issue of leg pains. A questionnaire was devised by one of the authors (S.H.) in order to gain insight into the prevalence of their concerns. Participation was offered to all members (3200) of the Little Hearts Matter support group through their social media sites and disseminated to other congenital cardiac charities. The study was also publicised in paediatric outpatient settings, allowing others with CHD, who were not members of Little Hearts Matter, to participate.

Survey design

The survey contained questions on general demographic information, diagnosis, medications, comorbidities, diet, and activity. Specific questions addressed the nature of leg pains and

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exacerbating and relieving factors. Questions were also asked regarding siblings, including whether they experienced leg pains. The survey questions are available in the Supplementary Table 1. The survey was performed using the SurveyMonkey web tool (https://www.surveymonkey.net) and results were collated by S.H. before being passed in fully anonymised form to the remaining authors for analysis.

Ethical approval

As the survey was initiated, developed, and administered by the Little Hearts Matter patient group (S.H.) and analysis was limited to data provided without patient-identifiable information, ethical approval was not required. This study was reviewed and the non-requirement for approval by a National Health Service Research Ethics Committee was confirmed by the chair of North East – Tyne & Wear South Research Ethics committee.

Statistics

Data are presented numerically and also as a percentage of the stated denominator. Associations were tested using χ^2 or Fisher's exact tests. When assessing the association between precipitating factors and methods of alleviating pain, p values were corrected using the sequential Bonferroni method and ϕ reported to reflect the degree of association. Data were analysed in SPSSv.24, with $p < 0.05$ considered statistically significant.

Results

A total of 220 respondents, of whom 57.4% were male, with a mean age of 8.3 years (0.2–29.8), completed the survey. Although parents and carers often completed the survey on behalf of the patient, the subject of the survey is described as the respondent in the resulting analysis. Demographic and diagnostic details are shown in Table 1.

The response rate was >90% for 14/16 questions, and all questions had a response rate >80%. The survey was available for 1 year; however, the majority of responses occurred within the first 3 months.

Description and impact of leg pains

In all, 206/220 (93.6%) survey respondents reported leg pain. The frequency of leg pains and precipitating factors are shown in Figures 1 and 2a. Each respondent identified a median of three out of nine precipitatory factors, with an inter-quartile range of zero to four). The majority of affected respondents experienced leg pains nocturnally (169/206, 82.0%) – a feature consistent with "growing pains" of childhood.[6] Other features consistent with growing pains were an association with day-time activity and an intermittent course. The location of the pain was typically bilateral and involved the lower legs including knees, shins, and muscles, but not thighs, hips, or toes. A range of phrases were used by respondents to describe the pains, including "cramps", "tightening", or "aches". Importantly, however, respondents mainly used the word "pain" itself to describe their symptoms, with just under half (101/206, 49.0%) reporting associated crying or screaming.

The methods used to alleviate pains are depicted in Figure 2b: analgesia (132/206, 64.1%), massage (111/206, 53.9%), and rest (107/206, 51.2%) were the most frequently used options. Improvement with resting was significantly associated with pain occurring after walking ($\phi = 0.277$), brought on by stress ($\phi = 0.260$), and with pain occurring while unwell ($\phi = 0.279$). Relief with elevation ($\phi = 0.248$) was also associated with pain brought on by stress, whereas relief with warming was associated with pain occurring in response to cold ($\phi = 0.294$). It is interesting that the strongest significant association was seen between nocturnal pain and relief with painkillers ($\phi = 0.335$). Nocturnal pain was also associated with relief with massage ($\phi = 0.276$).

Respondents also described their attempts to seek validation of these symptoms and the lack of available information to them. The nature and impact of leg pains is reflected in free text comments made by individual respondents (Table 2). A small number of respondents (8/206, 38.8%) described organic features of pain including limping, knee swelling, and unilateral symptoms. A total of 170 respondents reported having siblings, although only 107 (62.9%) provided information as to the presence or absence of leg pains. Leg pains were more frequently reported in respondents than their generally healthy siblings (32/107, 29.9%, $p < 0.001$).

Associations of leg pains with other factors

There was no gender bias in the reporting of leg pains – 94.6% were females and 92.9% were male, with $p = 0.60$ – or the frequency at which they occurred ($p = 0.57$). Although there was no relationship between reported leg pain and age, older respondents were more likely to associate leg pains with times when they felt stressed ($p = 0.02$) and at night ($p = 0.05$). Frequency of pains did not differ significantly between groups ($p = 0.28$). Individuals taking aspirin were more likely to report leg pains (98.2 versus 89.0%; $p = 0.01$). In all, 48% (105/220) of respondents described themselves as active either outdoors or indoors, whereas 51.8% (114/220) reported reduced levels of activity, with five being wheelchair-bound. Those who were more active were more likely to have frequent leg pains – more than once a week (50.0 versus 37.1%, $p = 0.04$).

In all, 69/220 (31.4%) described hypermobility, postural, or orthopaedic defects – including flat feet, high arches, talipes, hypochondroplasia, or scoliosis. All respondents with hypermobility ($n = 12$) reported leg pains. Individuals with hypermobility reported pain predominantly after exercise as is typical in this condition ($n = 9$, $p = 0.011$), and in hot weather ($p = 0.019$). Leg pains were not more prevalent in those with orthopaedic or postural issues.

Discussion

This is the first study to report leg pains in children and young people with CHD. Using an online survey, 93.6% respondents reported having leg pains, which was significantly higher than that reported in their generally healthy siblings (29.9%). The majority experienced pains nocturnally or when tired, and most of them alleviated symptoms with painkillers or massage.

Although reported pains are likely to reflect a mixture of aetiologies, the dominant features, such as nocturnal occurrence, intermittent nature, and focus in knees and calves, share clear similarities with benign childhood "growing pains".[6] "Growing pains" are a clinical diagnosis made when other potentially serious causes are excluded. Persistence of symptoms, presence of joint involvement, systemic features, limping, or abnormalities on examination or investigation should alert the clinician to other diagnoses.[7] Although the condition is self-limiting and the acknowledged prognosis benign, symptoms can be highly

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Table 1. Demographic details, diagnosis, comorbidities, and medications.

	All (n = 220)	Single-ventricle circulation (n = 153)	Biventricular circulation (n = 51)	Other (n = 16)
Male	126 (57.2%)	91 (59.5%)	30 (57.7%)	5 (31.3%)
Age (years)	8.3 (0.2–29.8)	8.7 (0.2–29.8)	6.6 (1.2–25.3)	7.6 (1.0–13.4)
Hypoplastic left heart		72	0	0
Hypoplastic right heart*		51	0	0
Double-inlet left ventricle		12	0	0
Mitral atresia or Shone's complex		5	1	0
Complete atrioventricular septal defect		4	3	0
Transposition of great arteries**		4	9	0
Truncus arteriosus		0	2	0
Tetralogy of fallot or variant		0	13	0
Aortic valve disease/coarctation		0	10	0
Other		0	13	2***
Unspecified		5	0	14
Fontan circulation	126 (57.2%)	126 (82.4%)	0 (0.0%)	0 (0.0%)
Biventricular Repair	45 (20.5%)	0 (0.0%)	45 (88.2%)	Unknown
Chromosomal abnormality		2	2	0
Pacemakers		7	2	0
Kidney or bowel comorbidity		14	6	0
Neurological comorbidity		10	0	0
Hypermobility	12 (5.5%)	8	4	0
Postural/orthopaedic condition	57 (25.9%)	36	18	3
Developmental or behavioural condition	10 (4.5%)	9	1	0
Aspirin	100			
Warfarin	60			
Beta-blockers	6			
ACE-I/ARB	85			
Inhalers	11			
Diuretics	14			

ACE-I = angiotensin converting enzyme inhibitor; ARB = angiotensin receptor blocker
*Including tricuspid atresia and pulmonary atresia
**Including simple and complex forms and congenitally corrected
***Including hypoplastic left heart syndrome with heart transplant, and Ebstein with Glenn shunt

distressing. The precise cause remains poorly understood. Various mechanisms are proposed and associations with obesity, orthopaedic factors such as flat feet, reduced bone strength, lowered pain thresholds, and psychosocial factors are documented.[8–13] Positive family history and suggested overlap with restless legs syndrome also indicate a possible genetic component.[10,14] Treatment with vitamin D supplementation, in a small Italian cohort study of healthy 5–11-year-olds, and efficacy of a twice-daily muscle stretching programme, in a small Canadian unblinded randomised controlled trial in 5–14-year-old healthy children, have suggested benefit in relieving symptoms.[15,16]

In our selected cohort with CHD, leg pains occurred more often than in their generally healthy siblings. Skeletal muscle and bone deficits, as well as vitamin D deficiency, have been observed in patients post Fontan completion, a dominant component group among respondents, and may therefore be implicated in the aetiology of their leg pains.[17,18] Patients with Fontan completion understandably report reduced quality of life; however, little work has been done to elucidate the effects of this on pain thresholds and reporting of symptoms.[19] Progress in CHD has been measured by stepwise improvements in survival. At present, most children born with these conditions are expected to survive into adulthood, and

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focus has shifted to functional capacity, morbidity, and quality of life as outcome measures. Although questionnaires such as PEDSQL, PCQLI, SF 36, and ConQol are validated and may provide useful information, these mainly explore areas defined by clinicians, although PCQLI does include some patient-defined measures.[20–22] Patients use social media and join online health communities to complement rather than bypass healthcare professionals, fulfilling unmet needs including informational and network support, as well as emotional expression or social comparison.[3,23] The relationship with the healthcare professional is generally viewed by patients as a clinical one, where the professional provides expertise and treatment based on medical knowledge, but not first-hand experience.[23,24] Although patient groups can offer practical and emotional support between patients with similar experience, which is more difficult to provide in the traditional medical setting, support may be limited if problems and potential solutions are not recognised and addressed by clinicians.

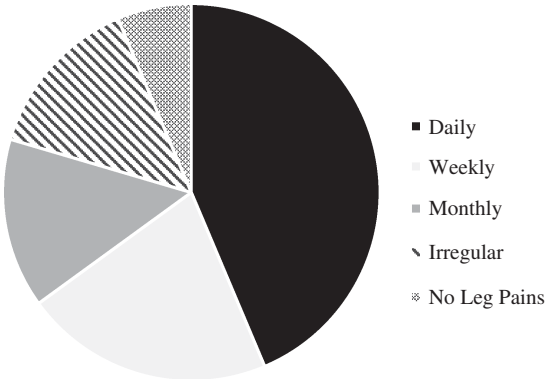


Figure 1. Frequency of leg pain in respondents.

In this study, the poorly documented yet clearly distressing symptom of leg pain was identified by Little Hearts Matter from their patient base, and a larger survey was undertaken. Although limitations (see below) exist for this type of data, the results can direct researchers to investigate new areas of unmet need, which may be of growing importance to patients as issues of mortality and morbidity are addressed. However, focused efforts are required to validate patient-reported outcome tools relevant to

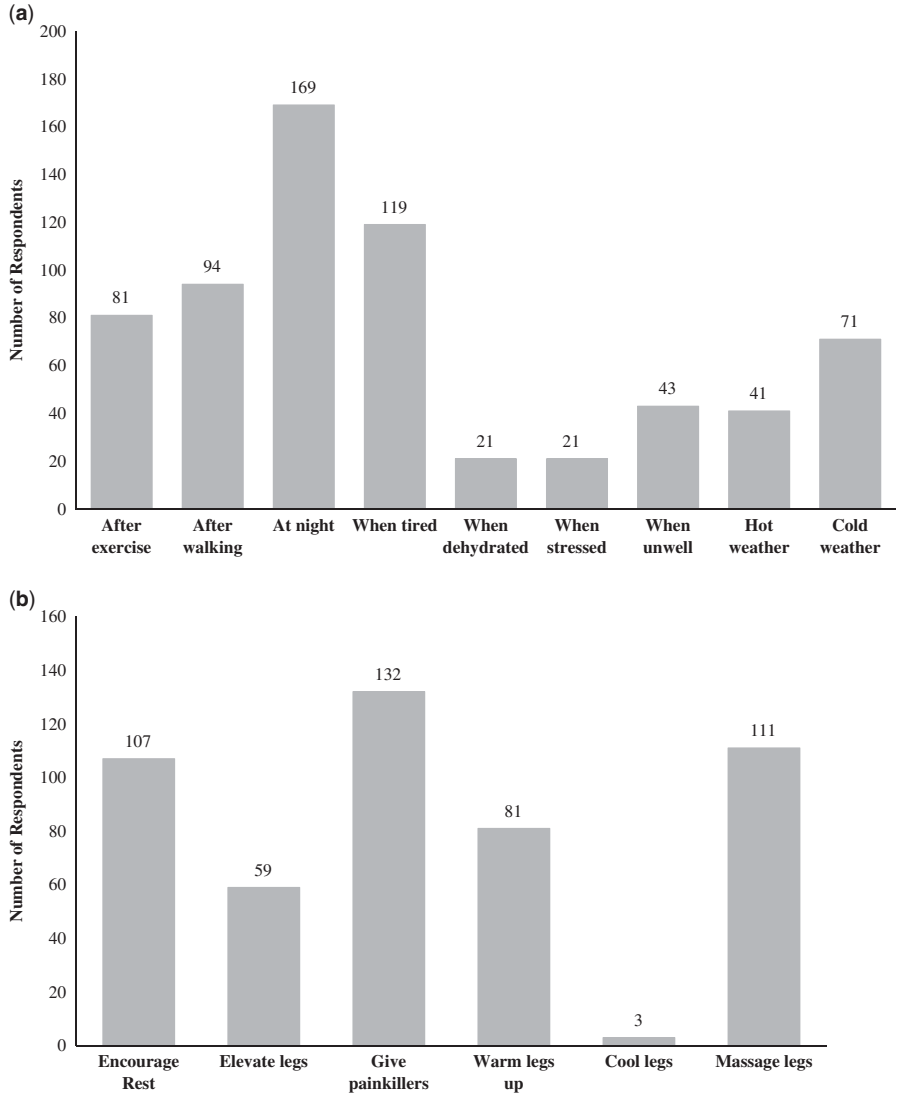


Figure 2. (*a*) Factors precipitating leg pains and (*b*) methods used to alleviate leg pains.

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Table 2. Respondent comments reflecting nature of leg pains and their impact on the patient and family.

Respondent comments	
Nature of pains	• Described as "cramps", "tightening", or "aches"
Intermittent course	• "It happens in waves. They hurt for a while about 6 months ago and it was an everyday pain. Then it stopped for a few months, and about 2 months ago she said they were hurting again. At the current time, they aren't bothering her"
Relationship with activity	• "If X has a very active day e.g. sports day or a day with not much rest, then leg pain will occur that night, sometimes in the day but that is rarely"
Distressing nature	• X doesn't really cry if he hurts himself and is quite tough but this leg pain will always bring him to tears and he will be holding the leg around the knee and is very restless. He finds it difficult then to get comfortable" • "She is inconsolable at night time. It's like she's panicking, she can't keep still and settle to sleep"
Attempts to seek validation	• "My 17-year old son has been complaining of leg pain for years. Just last month, we spoke to his cardiologist who couldn't identify with it from a heart perspective" • "She wakes up crying (has never managed a full night if sleep!) and quite frequently she tells us that she has pains in her legs. We have mentioned this to her consultant"
An unmet need?	• "I would certainly have liked some more information about it as at the time we thought it was just him"

this population to facilitate this. Better education of CHD healthcare professionals in the patient experience could also help alleviate perceived contradictions with scientific knowledge, which in other domains has been a recognised source of distress to patients, resulting in detrimental consequences.[25]

This study suffers from reporting bias, as it is likely that individuals with leg pains were more likely to respond to a questionnaire on this subject than those without. Parents and carers responding on behalf of their children may also influence reporting rate. Although we cannot provide exact numbers, it is apparent that around 90% of respondents were parents or carers responding on behalf of their child. Discrepancies in patient and parent reporting of quality-of-life issues are recognised and may reflect differences in illness perception.[26] It is interesting that in a few cases parents reported leg pains in young babies, where it may be difficult to conceive how this symptom could be expressed. This may reflect unaddressed needs in the carers, as well as the physical symptoms of the child. Nevertheless, the difference seen between patients and their healthy siblings, whose rate of leg pains was consistent with that reported in normal children, suggests that this condition is more common in those with CHD, although admittedly the age of the siblings is unknown and sibling data were incomplete.[7] The overall response rate as a proportion of Little Hearts Matter members may be interpreted to be low, but it is likely that not all Little Hearts Matter members engage with the social media sites through which it was publicised. Further, because of the methodology used, it is not possible to be completely certain how many potential respondents there were. It would be important to repeat the survey, or a modified version, in another CHD population to validate the results. However, nearly everyone had leg pains, and thus there was low statistical power to analyse associations. In addition, we tested multiple associations in this study, and therefore there is an increased risk of type I errors. It is evident, for example, that participants report several precipitatory factors each for their leg pains; elucidating the contribution of each is likely to require both more detailed questioning and larger sample populations than available in the present study.

It is also the case that not all leg pains should necessarily be attributed to "growing pains", as this was not a validated questionnaire, rather a survey designed by a patient group to address their members' needs. Other limitations to the results reported are that comorbidities in this population are likely to be underreported as, although members of this group are typically well-educated about their condition, lay understanding or description of medical terms may be restricted. Equally, a degree of subjective interpretation has had to be applied by the investigators to permit the responses to be analysed using statistical methods. In addition, more refined open questions could have enhanced the richness of the qualitative data set obtained with respect to the nature of the leg pains.

Leg pains are an under-recognised, distressing symptom for patients with CHD and are reported more frequently than in their generally healthy siblings. It would be interesting to know how leg pains have an impact on the quality of life, participation in sports and school, and to understand whether and how medical and surgical intervention may influence their manifestation. This requires further investigation and offers an important example of how patient experiential knowledge can identify new areas for research and thus address perceived inconsistencies with existing medical knowledge.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S104795111800094X

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Conflicts of Interest. Dr David Crossland is a Medical Advisor to Little Hearts Matter.

Ethical Standards. Analysis of data was carried out by Newcastle University as a contracted service provider on behalf of the charity Little Hearts Matter. All data shared were fully anonymised. These arrangements were approved by the Chairman of North East – Tyne & Wear South NHS REC and further ethical approval was not required.

References

1. Ronksley PE, Sanmartin C, Quan H, et al. Association between chronic conditions and perceived unmet health care needs. Open Med 2012; 6: e48–e58.
2. Ronksley PE, Sanmartin C, Quan H, et al. Association between perceived unmet health care needs and risk of adverse health outcomes among patients with chronic medical conditions. Open Med 2013; 7: e21–e30.
3. Kirk S, Milnes L. An exploration of how young people and parents use online support in the context of living with cystic fibrosis. Health Expect 2016; 19: 309–321.

4. George DR, Rovniak LS, Kraschnewski JL. Dangers and opportunities for social media in medicine. Clin Obstet Gynecol 2013; 56: 453–462.
5. Hall S, Chen S, Kao A, et al. 162 symptom impact and unmet need in systemic/cutaneous lupus erythematosus: results from a patient-centred study set in a social media community. Lupus Sci Med 2017; 4: A77.
6. Peterson H. Growing pains. Pediatr Clin North Am 1986; 33: 1365–1372.
7. Mohanta MP. Growing pains: practitioners' dilemma. Indian Pediatr 2014; 51: 379–383.
8. Hawksley JC. Growing pains in relation to rheumatism. Br Med J 1939; 1: 155–157.
9. Evans AM, Scutter SD. Are foot posture and functional health different in children with growing pains? Pediatr Int 2007; 49: 991–996.
10. Evans AM, Scutter SD, Lang LMG, Dansie BR. "Growing pains" in young children: a study of the profile, experiences and quality of life issues of four to six year old children with recurrent leg pain. Foot 2006; 16: 120–124.
11. Friedland O, Hashkes PJ, Jaber L, et al. Decreased bone speed of sound in children with growing pains measured by quantitative ultrasound. J Rheumatol 2005; 32: 1354–1357.
12. Hashkes PJ, Friedland O, Jaber L, Cohen HA, Wolach B, Uziel Y. Decreased pain threshold in children with growing pains. J Rheumatol 2004; 31: 610–613.
13. Naish JM, Apley J. "Growing pains": a clinical study of non-arthritic limb pains in children. Arch Dis Child 1951; 26: 134–140.
14. Walters AS, Gabelia D, Frauscher B. Restless legs syndrome (Willis–Ekbom disease) and growing pains: are they the same thing? A side-by-side comparison of the diagnostic criteria for both and recommendations for future research. Sleep Med 2013; 14: 1247–1252.
15. Morandi G, Maines E, Piona C, et al. Significant association among growing pains, vitamin D supplementation, and bone mineral status: results from a pilot cohort study. J Bone Miner Metab 2015; 33: 201–206.
16. Baxter MP, Dulberg C. "Growing pains" in childhood – a proposal for treatment. J Pediatr Orthop n.d.; 8: 402–406.
17. Avitabile CM, Leonard MB, Zemel BS, et al. Lean mass deficits, vitamin D status and exercise capacity in children and young adults after Fontan palliation. Heart 2014; 100: 1702–1707.
18. Avitabile CM, Goldberg DJ, Zemel BS, et al. Deficits in bone density and structure in children and young adults following Fontan palliation. Bone 2015; 77: 12–16.
19. Uzark K, Zak V, Shrader P, et al. Assessment of quality of life in young patients with single ventricle after the Fontan operation. J Pediatr 2016; 170: 166–172.e1.
20. Macran S, Birks Y, Parsons J, et al. The development of a new measure of quality of life for children with congenital cardiac disease. Cardiol Young 2006; 16: 165–172.
21. Idorn L, Jensen AS, Juul K, et al. Quality of life and cognitive function in Fontan patients, a population-based study. Int J Cardiol 2013; 168: 3230–3235.
22. Marino BS, Shera D, Wernovsky G, et al. The development of the pediatric cardiac quality of life inventory: a quality of life measure for children and adolescents with heart disease. Qual Life Res 2008; 17: 613–626.
23. Smailhodzic E, Hooijsma W, Boonstra A, Langley DJ. Social media use in healthcare: a systematic review of effects on patients and on their relationship with healthcare professionals. BMC Health Serv Res 2016; 16: 442.
24. Colineau N, Paris C. Talking about your health to strangers: understanding the use of online social networks by patients. New Rev Hypermedia Multimed 2010; 16: 141–160.
25. Koschack J, Weibezahl L, Friede T, Himmel W, Makedonski P, Grabowski J. Scientific versus experiential evidence: discourse analysis of the chronic cerebrospinal venous insufficiency debate in a multiple sclerosis forum. J Med Internet Res 2015; 17: e159.
26. Eiser C, Varni JW. Health-related quality of life and symptom reporting: similarities and differences between children and their parents. Eur J Pediatr 2013; 172: 1299–1304.

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