

Parental perceptions of bladder dysfunction in children with symptomatic joint hypermobility.

Authors: Marianna Veriki^{1,2}, Annette Coomer^{1,3}, Jane Simmonds^{1,4}

- 1. UCL Great Ormond Street Institute of Child Health, University College London, UK*
- 2. Children's Physiotherapy Homerton University Hospital, London, UK*
- 3. Children's Physiotherapy, Imperial College Healthcare, NHS Trust, London, UK*
- 4. London Hypermobility Unit, Central Health Physiotherapy, Wellington Hospital, London UK*

Abstract

Children with symptomatic joint hypermobility (SJH) report chronic pain, fatigue and joint instability as their main symptoms. Symptoms can extend beyond musculoskeletal and include lower urinary tract dysfunction (LUTD). There are no studies exploring parent-perceived Quality of Life (QoL) in children with LUTD, or strategies used to manage symptoms in this population.

Our online cross sectional survey assessed parents of 6 to 18 year olds via two UK charitable organisations. It included the Paediatric Incontinence Questionnaire (PinQ), the Dysfunctional Voiding Symptom Score (DVSS), and questions exploring treatments and efficacy.

Sixty-seven parental questionnaires were analysed. The majority of children were white females (95%, n= 64). Urgency (97%, n=65) and enuresis (41.7%, n=28) were the most frequently reported symptoms. The PinQ questionnaire showed that children were emotionally distressed by LUTD. Twenty nine parents (43%, n=29) reported pharmacotherapy as the most effective treatment. Few had accessed other treatments such as physiotherapy (16%, n=18). Most parents reported that overall treatment did not meet their expectations.

This study highlights that parents perceive LUTD as having a great impact on their child's QoL. There is need for education amongst clinicians about LUTD and hypermobility-related disorders, and for development and evaluation of treatments.

INTRODUCTION

Joint hypermobility (JH) is a term that is universally accepted to describe the capability of a joint to move beyond normal limits along the physiological axes and is dependent on age, gender and ethnicity (Castori et al., 2017). Castori et al (2017) suggested a classification of JH where asymptomatic JH, Hypermobility Spectrum Disorders (HSD), and hypermobile Ehlers-Danlos Syndrome (hEDS) are part of the spectrum of the more common hypermobility-related disorders. For the purposes of this study, symptomatic JH (SJH) will be used as an umbrella term to exclude patients with asymptomatic JH while describing patients within the spectrum passing through HSD and hEDS.

Recent research in individuals with SJH revealed that symptoms extend beyond the musculoskeletal system (de Kort et al., 2003; Gazit et al., 2016). An increasing number of studies provide robust evidence on the presence of lower urinary tract dysfunction (LUTD) as part of the clinical presentation of SJH (de Kort et al., 2003; Engelbert et al., 2017). Numerous authors have identified a correlation between urinary incontinence and collagen abnormalities (de Kort et al., 2003; Kajbafzadeh et al., 2014; Ulmsten et al., 1987) and suggest that these abnormalities can cause a dyssynergic function of their pelvic floor musculature (PFM) (Junginger et al., 2014; Mastoroudes et al., 2013).

Evidence to support the most effective treatment of LUTD for the paediatric population is limited. Initial approaches to management include urotherapy which consists of education on voiding position, fluid management and timed voiding (Austin et al., 2011). Other treatments include pharmacotherapy, pelvic floor muscle training (PFMT) with or without biofeedback and surgical interventions for progressive cases (Sureshkumar et al., 2003). Currently, there is no standardised protocol for the treatment of children with LUTD, apart from guidelines for bed time wetting (Evans et al., 2010). As parents are the gatekeepers of their child's care it would be meaningful to evaluate whether LUTD treatment options commonly reported in literature and used in clinical practice are endorsed by parents. Moreover, no studies have been conducted that explore the management of LUTD in children with SJH.

LUTD, if not managed properly, can have a significant impact on a child's life (Thibodeau et al., 2013, Marciano et al., 2018, Bower et al., 2006, Deshpande et al., 2011). Marciano et al (2018) found that self-esteem and mental health were the most affected domains. The clinical significance of LUTD and its effect on QoL in this group of children with SJH remains unclear.

The aim of this study is to explore the parental perceptions of children with SJH in relation to the symptoms of LUTD, their QoL and the effectiveness of treatments they have sought.

METHODS

Study design and data collection

A cross-sectional survey using an online questionnaire was conducted to explore the perceptions of parents on their child's LUTD and the treatment options they sought. The study was approved by the University College of London (UCL) research ethics committee (number:14723/001).

The sample population were parents who are members of the Hypermobility Syndromes Association (HMSA) and the Ehlers-Danlos Support UK (EDSUK). Participants were English speaking parents of children aged 5-18 living in the UK. Children with a history of known pathological, neurological, rheumatological condition, cognitive illness, neurogenic bladder or known anatomical abnormality of the lower urinary tract were excluded.

Participants were recruited via an advertisement on the charities' official websites which guided them to an online survey platform.

Online Questionnaire

The questionnaire was delivered through the online survey tool "Survey Monkey" (<https://www.surveymonkey.com/>) and consisted of four domains:

- 1) Demographics: information about their child's age, gender and ethnicity.
- 2) Diagnosis and clinical presentation of SJH and LUTD. A modified version of the Dysfunctional Voiding Symptom Score (DVSS) was used to identify symptoms of LUTD.
- 3) Quality of life through the Paediatric Incontinence Questionnaire (PinQ) (Bower et al., 2006).
- 4) Treatment options and perceived effectiveness.

Additional open questions were included, which addressed parents' perception of the treatment effectiveness and provided opportunity for additional feedback.

Data Analysis

Prior to data collection, a pilot study was conducted to test the face validity, coding system, collection and processing of the data. The questionnaire was piloted with supervisors, parents, expert clinicians and researchers specialising in hypermobility syndromes.

Descriptive analysis was conducted for demographic data, presenting symptoms and beliefs data to determine population characteristics and perceptions on QoL, choice of LUTD treatments and perceived effectiveness. Responses to open questions were analysed by using a content analysis as described by Cohen (1960) and Graneheim et al (2003).

RESULTS

Demographics

Out of 133 participants who accessed the survey, data from 67 participants were deemed complete and included for analysis. Forty-eight percent (48%, n=32/67) of children were white females diagnosed with hEDS, as reported by their parents. More characteristics of children represented by their parents are shown in Table 1.

Symptoms of SJH and LUTD

The majority of patients with moderate SJH (53%, n=36) also presented with moderate pain (44.8%, n=30/36), intermittent type of pain (43.3%, n=29/36) and four or more painful joints or areas (77.7%, n=28/36).

Based on the inclusion criteria for this study, LUTD symptoms were present in all children represented by their parents. Although 97% (n=62) of parents reported the presence of urgency as one of their child's LUTD symptoms, only half of them (49.2%, n=33) reported that it occurs 'almost every time'. The least reported symptom was pain during voiding which was reported to occur 'less than half of the time' in 74.6 % (n=50) of parents (Figure 1).

Quality of life

Figure 2 shows parents' responses to the adapted version of PinQ assessing variables of QoL. Results showed that the majority of affected factors in a child's QoL are 'almost always' or 'sometimes' thought to be related to intrinsic factors. Participation in activities varied in frequency of occurrence (Figure 2).

Perceived treatment effectiveness

All parents (n=67) had sought treatment for their child's LUTD. Seventy-percent of parents (n=47) found that regular toilet trips, as a strategy, were 'moderately' to 'very' effective for their child's LUTD. Parents also found that pharmacotherapy, education of toilet position, waking up to void and use of protective underwear were 'moderately' to 'very' effective (59.7%, n=40, 40.3%, n=27, 38.8%, n=26, 29.9%, n=20 respectively). Figure 3 shows a summary of these results and highlights that the majority of conservative treatments were 'not tried' by parents. A great number of parents (29%, n=18) rated pharmacotherapy as the single most effective treatment for their child's LUTD, followed by scheduled voiding (12.9%, n=8). Interestingly, thirteen parents (19.4%) reported that nothing was effective for their child's LUTD.

Open questions

Not all parents answered the open questions. More than half of the parents reported that their expectations in relation to the treatment they chose were not met (58%, n= 29/50). Parents frequently disclosed their disappointment with their health care provider (HCP) (54%, n=20/37), which was often related to the lack of support or inadequate information provided by the HCPs in relation to the link between SJH and bladder problems (43.2%, n= 16/37). When parents were asked which treatment they would choose if their symptoms persisted, 29% reported that they would choose medication, but an equal proportion of parents reported that they were not sure of which treatment they would choose in the future (Table 2). Although medication was reported as one the preferred future treatments, some parents expressed their wish to withdraw from medication mainly due to the associated side effects.

DISCUSSION

This research explored parental perceptions of treatments for LUTD) and provided an understanding of the domains affected in the life of children with SJH who also encounter lower urinary tract problems.

Quality of life

Studies on children with LUTD have shown no statistically significant difference between parent and child responses on the PinQ, highlighting that parents can reliably report the impact of LUTD on their child's QoL (Thibodeau et al., 2013, Marciano et al., 2018). Thibodeau et al (2013) found a positive correlation between symptom severity scores (DVSS scores) and PinQ scores in children with LUTD, however, the most affected domains were not demonstrated. In this study, the intrinsic domains of life were mostly affected, and a high percentage of parents reported that their child was missing activities as a result of their LUTD (e.g. missing sleepovers, visits toilet while watching a movie). Similar findings were reported by Pacey et al (2015) who found that stress incontinence was the most distressing factor in the life of 23 children with SJH.

Treatments of LUTD

Medication was perceived as moderately to very effective by approximately 60% (n=40) of parents. Considering the extensive use of medication in practice (van Engelenburg–van Lonkhuyzen et al., 2017), it is surprising that only 29.1% of parents would use pharmacotherapy if their child's symptoms persisted. Uptake of and compliance to medication may be limited in this group, as it is noted that families are more likely to adhere to the treatments that they perceive as effective (Claar & Scharff, 2007).

Conservative options were not frequently employed by parents. Specifically, less than half of the parents (48.3%, n=32) perceived urotherapy as an effective management option and none would choose it in the future if symptoms persisted, despite the recommendations from the International Children's Continence Society (ICCS) (Chase et al., 2010).

The issues of bed lifting in relation to postponement of dryness have been discussed in literature since 1982 (Christmanson and Lisper, 1982) and suggest that this strategy will not promote long-term dryness (NICE, 2010) . However, results from this study show that parents still use it as a means to manage their child's enuresis (38.8%, n=26), despite the absence of

bed lifting as a recommended strategy by the National Institute for Health and Care Excellence (NICE) guidelines (Evans et al., 2010).

Although management of LUTD with pelvic health physiotherapy in adults has shown to be effective (Hay-Smith & Dumoulin, 2006), there is little evidence of physiotherapy as an intervention for paediatric LUTD which may make doctors and HCPs reluctant to recommend it. In addition, the role of physiotherapy or PFMT is not reported in paediatric continence guidelines (Evans et al., 2010) which may explain why PFMT and physiotherapy were not tried by 76.1 and 83.6% of parents, respectively.

Gaps in current service provision

Positive health outcomes are associated with patient satisfaction (Hush et al., 2012). In this study, disappointment in HCPs was reported by the majority of parents and may represent a possible pitfall in current service provision. Similar inferences can be drawn by the respondents who reported they were not aware of the link between SJH and LUTD and that they received inadequate information by their HCPs. The majority of physiotherapists are not familiar with the extra articular features of SJH (Billings et al., 2015). This is likely to be due to limited evidence and lack of standardised guidelines on the systemic manifestations of SJH and their management.

Limitations

Replication of the study with a greater sample size would provide a better representation of the population. In addition, it is likely that theme saturation from parents' responses to the open questions was not achieved. The total number of parents who answered the survey was 133, yet only 58% of the questionnaire was completed by the majority of the participants.

Another limitation to this study is that it used adapted versions of recognised outcome measures and therefore the total scores for these components could not be calculated and analysed. This compromised the comparison to other studies.

Recruitment of the participants from the HMSA and EDSUK may have resulted in bias. Members of these charities may be those who have already been diagnosed with SJH who may not represent the wider demographics and reduce the generalisability of the current findings.

CONCLUSION

In this research, parents perceived LUTD to markedly impact on several QoL domains in children with SJH. Clinicians are therefore encouraged to firstly assess possible LUTD in children attending clinics and to explore QoL domains in order to target interventions.

The treatments perceived most effective by parents are recommended by the current NICE guidelines for clinical practice, though the implementation of more conservative options as first-line treatments could be considered in clinical practice. Future research should address the efficacy of conservative interventions for children with SJH and LUTD.

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TABLES AND GRAPHS

Table 1- Demographics of children who were represented by their parents.

Characteristics		Total % (n)
	Male	31.3 (21)
	Female	68.7 (46)
	Age*	*10.8± 4.09
Ethnicity	White	95.5 (64)
	Mixed	4.5 (3)
Diagnosis	Joint hypermobility with symptoms	13.4 (9)
	Hypermobile EDS	47.8 (32)
	Joint hypermobility syndrome	38.8 (26)
	Presence of LUTD	100 (67)
	Chronic LUTD	97 (65)
	Age of LUTD onset *	*3.69± 2.9
<p><i>KEY: SJH: Symptomatic Joint hypermobility, hEDS: Hypermobile Ehlers-Danlos Syndrome, LUTD: Lower urinary tract dysfunction. Chronic LUTD: >3 months of symptoms *Normality observation showed that 'age' and 'age of LUTD onset' were normally distributed. Expressed in mean ±SD</i></p>		

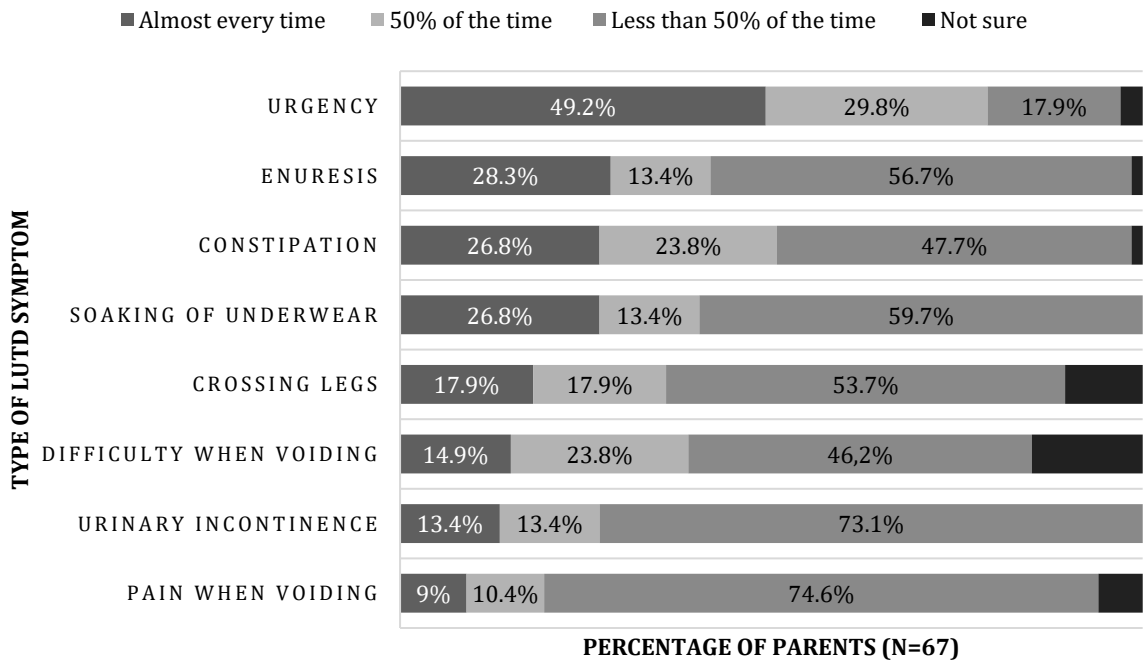


Figure 1- Percentage of presenting LUTD symptoms as reported by parents when answering the DVSS.

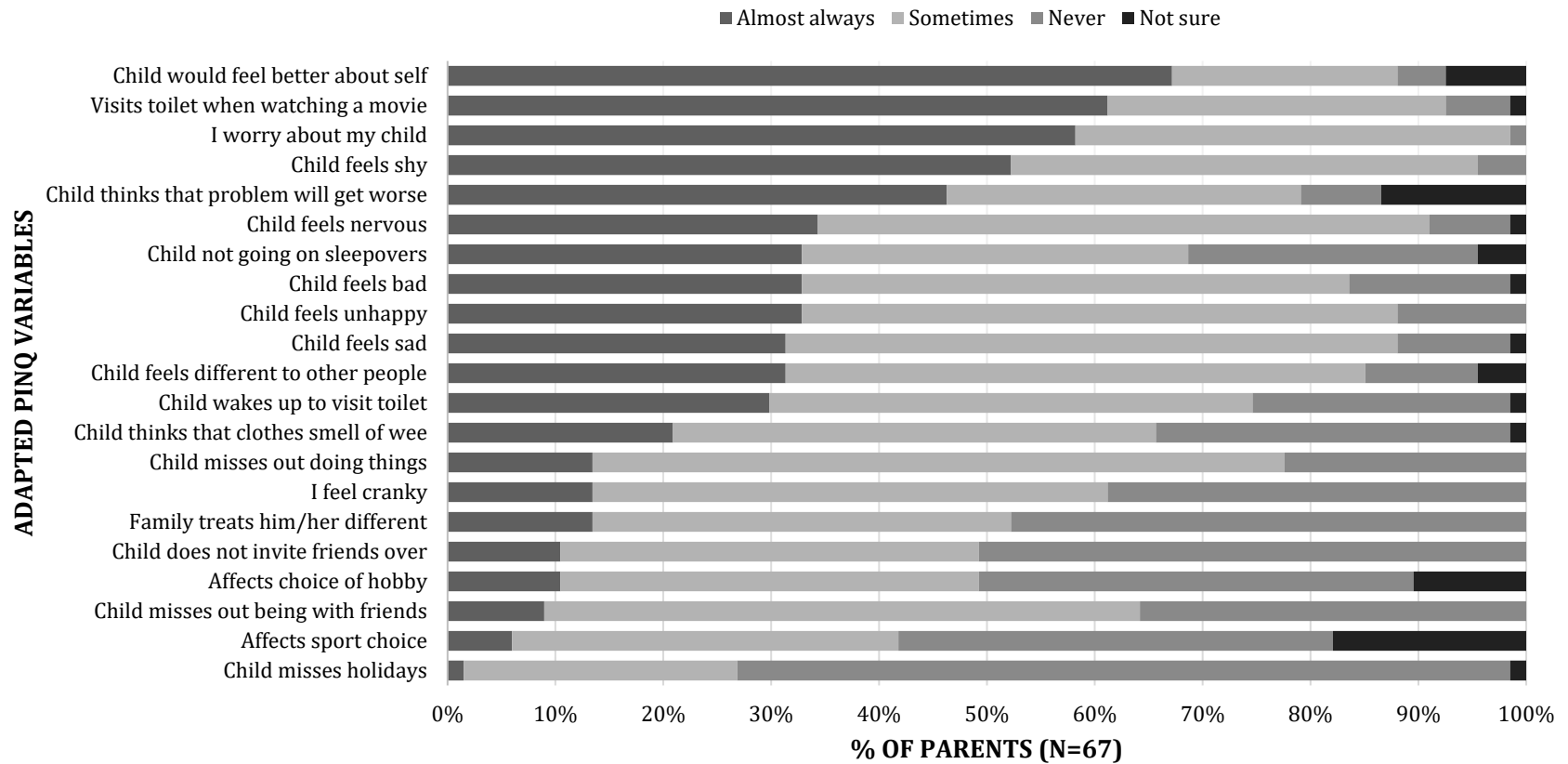


Figure 2- Percentages of parents reporting about their child's Qo;L by answering the adapted version of PinQ.

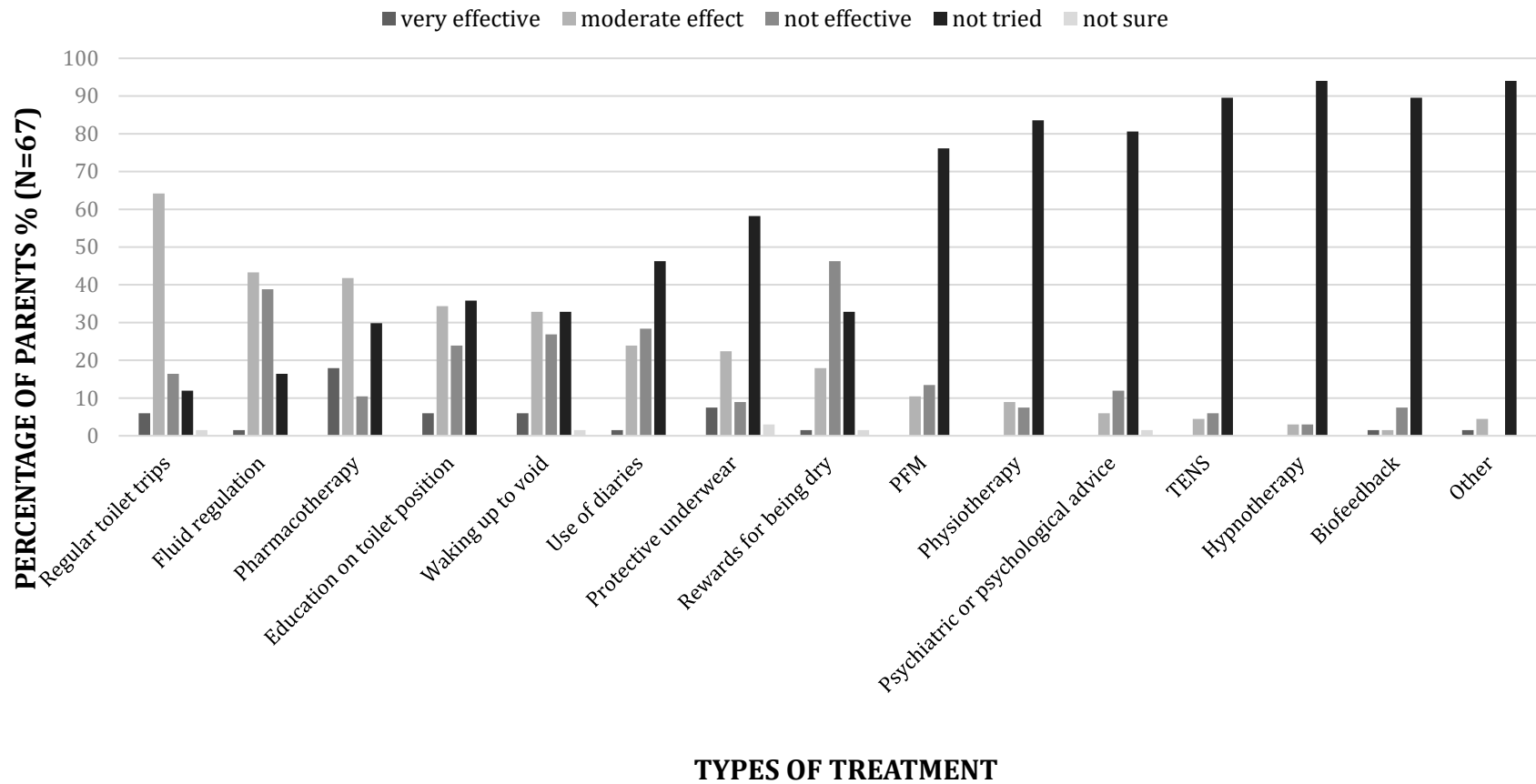


Figure 3- Common treatment options found in literature and how effective parents perceive them

Table 2- Frequency of parents reporting the treatment choice they would choose in the future.

Coded theme	Frequency	Valid percentage (%)
Not sure (Uncertainty)	14	29.1
Medication	14	29.1
Seek for further advice	6	12.5
Would try anything	4	8.3
Bed wetting alarm	3	6.2
Other*	7	14.5
TOTAL	48	100
<i>KEY: *included Botox and surgery, PFMT: Pelvic floor muscle training, physiotherapy</i>		

