The Standardization of Terminology of Bladder Function in Children and Adolescents: **Update Report from the Standardization Committee of the International Children's Continence Society (ICCS)**

Paul F. Austin*, Stu Bauer, Wendy Bower, Janet Chase, Israel Franco, Piet Hoebeke, Søren Rittig, Johan Vande Walle, Alexander von Gontard, Anne Wright, Stephen S. Yang and Tryggve Nevéus

From the Division of Urology, Washington University in St. Louis, St. Louis Children's Hopsital,USA (PFA), Department of Urology, Children's Hospital and Harvard Medical School, Boston, USA (SB), Pediatrics (Nephrology Section), Skejby University Hospital, Aarhus, Denmark (WB, SR), The Children's Centre, Cabrini Hospital, Melbourne, Australia (JC), New York Medical College, , USA (IF), Pediatric Urology and Nephrology, Gent University Hospital, Ghent, Belgium, (PH, JVdW), Department of Child and Adolescent Psychiatry, Saarland University Hospital, Germany (AvG), Pediatrics, Evelina Children's Hospital, St Thomas' Hospital, London, England (AW), Division of Urology, Tzu Chi Taipei Hospital, New Taipei and Buddhist Tzu Chi University, Hualien, Taiwan (SSY) and Section of Paediatric Nephrology, Department of Women's and Children's Health, Uppsala University, Uppsala, Sweden (TN).

*Corresponding Author:
Paul F. Austin
Washington University School of Medicine
4990 Children's Place
Suite 1120, Campus Box 8242
Pediatric Urology
Saint Louis, MO 63110-1077
Phone Number: 314-454-6034
E-mail Address: austinp@wustl.edu

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ABSTRACT

Purpose:
The impact from the original International Children's Continence Society (ICCS) terminology document on lower urinary tract (LUT) function resulted in the global establishment of uniformity and clarity in the characterization of LUT function and dysfunction in children among multiple healthcare disciplines. The present document serves as a stand-alone terminology update reflecting refinement and advancement of knowledge on pediatric LUT function.

Materials and Methods:
We assembled a variety of worldwide experts from multiple disciplines that care for children with LUT dysfunction were assembled. A critical review of the original ICCS terminology document and the current literature since its publication was performed. Additionally, contributions and feedback from the multidisciplinary ICCS membership were obtained.

Results:
A review of the literature over the last 7 years was performed. The ICCS experts assembled a new terminology document reflecting the current understanding of bladder function and LUT dysfunction in children using the resources from the literature review, expert opinion and ICCS member feedback.

Conclusions:
The present ICCS terminology document provides a current and consensus update to the evolving terminology and understanding of LUT function in children.

INTRODUCTION

The standardization of terminology for pediatric bladder and bowel function is critical to provide a platform for optimal understanding, communication and treatment among multiple health care providers who care for children and adolescents with LUT dysfunction. Terminology that is applicable internationally is particularly important because of the global prevalence of pediatric LUT dysfunction and the numerous specialists who treat these children and adolescents. LUT dysfunction is a broad term that encompasses subsets of LUT dysfunction with different manifestations. The heterogeneity of symptoms is at times overlapping and at other times unique to the subsets of LUT dysfunction. Universally accepted terms of pediatric LUT dysfunction is thus imperative to reduce confusion among care providers. Standardized terms are also critical for comparing research and study outcomes to optimally promote investigative understanding of pediatric LUT dysfunction.

The International Children's Continence Society (ICCS) is a unique organization whose members comprise multiple disciplines and specialties that care for children with bladder and bowel incontinence. Specialties include urology, nephrology, gastroenterology and psychiatry/psychology. Care providers include physicians, nurses, nurse practitioners, physiotherapists, psychologists, developmental pediatricians and physician assistants. The ICCS has regional representatives in every continent and has conducted numerous workshops and courses throughout the world. The ICCS is thus uniquely positioned to provide guidance in the standardization of terminology for BBD in children and adolescents.

The original report from the Standardization Committee of the International Children's Continence Society (ICCS)[1] provided new definitions and established a standardized terminology that has allowed clarity of communication over the last decade. The impact of the ICCS-proposed terminology on the body of literature of pediatric LUT function has been evaluated.[2] The importance of pediatric urinary incontinence is supported by the finding of a 49% increase in publications from 2002–2005 to 2007–2010 (55–82 per year). Additionally, there was approximately a fourfold increase in the likelihood of usage of ICCS recommended terminologies post ICCS guideline publication (OR: 4.19, 95% CI: 3.04–5.78, $P < 0.001$). It is noteworthy that there was no significant geographical variation in adoption of ICCS terminology. Despite the significant impact of the global adoption of ICCS terminology, approximately 25% of the studies published between 2007 and 2010 were noted to use obsolete terminologies.[2]

Similar to the dynamic flux of knowledge and understanding within medicine, the terminology for pediatric bladder and bowel function is dynamic. This document on the ICCS terminology of pediatric bladder and bowel function serves as a stand-alone terminology update reflecting refinement and advancement of knowledge on pediatric bladder and bowel function. Adherence to the updated terminology is followed by all ICCS courses and workshops and we encourage all investigators and clinicians that publish on the topic of pediatric bladder function to utilize the ICCS recommended terminology. To delineate

manuscripts and publications that follow the ICCS guidelines for the terminology of pediatric bladder and bowel function, we recommend future manuscripts include the text “Terminology adheres to the standards recommended by the International Children’s Continence Society except where specifically noted”.

MATERIALS AND METHODS

We assembled a distribution of worldwide experts from multiple disciplines that care for children with LUT dysfunction. Healthcare disciplines included urology, nephrology, general pediatrics, physical therapy and psychiatry. The panel of experts represented North America, Europe, Australia and Asia. A critical review of the original ICCS terminology document and the current literature was performed. Additionally, contributions and feedback from the multidisciplinary ICCS membership were obtained. Input from ICCS members include representation from North and South America, Europe, Middle East, Asia, Africa and Australia.

This terminology document represents the 3rd published standardization on terminology for lower urinary tract function and builds upon the previous ICCS documents[1, 3]. Recognition and reference to the terminology on lower urinary tract function by the International Continence Society (ICS)[4] as well the joint terminology for female pelvic floor dysfunction by the International Urogynecological Association (IUGA) and ICS[5] were employed. Additionally, we took into consideration the terms and definitions employed by the new Fifth Edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5).[6, 7]

There is no intent for this update document to serve as a guideline for clinical treatment. There are numerous previous ICCS documents outlining treatment for specific lower urinary tract conditions and associated co-morbid conditions.[8-15] This terminology update follows the prior ICCS terminology outline of establishing syntax to properly convey the symptoms of lower urinary tract dysfunction and to affirm terminology for investigative tools, signs, conditions and treatment parameters as they pertain to lower urinary tract function and dysfunction. We have updated the relevance of age to bladder and bowel function and discuss the commonality of bowel emptying issues with bladder function. We recognize that we are an organization that has primary expertise in bladder continence and function but equally recognize that there is a close relationship between bowel and bladder function. Thus, we emphasize the importance of bowel related terms in relation to bladder function as warranted.

TERMINOLOGY

Bladder and bowel dysfunction (BBD)

Because of the aforementioned relationship between the bladder and bowel we have termed concomitant bladder and bowel disturbances as BBD. We discourage the use of the term dysfunctional elimination syndrome (DES) as this connotes a particular abnormality or condition. We recommend BBD as a more descriptive comprehensive term of combined bladder and bowel disturbances that does not explain pathogenesis but is a term that is encompasses lower

urinary and bowel function. BBD is an umbrella term that can be subcategorized into lower urinary tract (LUT) dysfunction and bowel dysfunction. (Figure 1). When we use the term dysfunction or disorder, this represents clinical significance and relevance. In a research document or reference, authors should specify and provide support of using term BBD. In the absence of any co-morbid bowel dysfunction, then the term LUT dysfunction alone suffices.

SYMPTOMS

Symptoms are classified according to their relation to the storage and/or voiding phase of bladder function. Although a symptom may occur only once or rarely, this does not necessarily make it a condition. Symptoms are variable in LUT dysfunction and the duration of a symptom may alter the perception of its relevance.

The terminology used for LUT symptoms will focus on descriptive terms and avoid using quantitative language, as there is a lack of quantitative data to define symptomatic terms. Age of the child, however, has special relevance when applying terminology for pediatric bladder function. For function of the LUT, we will use 5.0 years of age as the reference for symptoms as this is the age used by the DSM-5 and the International Classification of Diseases-10 (ICD-10) to characterize urinary incontinence disorders.[7, 16] For functional bowel dysfunction the minimum age is 4.0 years. We recognize the variability and maturational aspect of LUT function[17] and fully acknowledge there are children who have voluntary control over LUT function < 5 years of age and therefore the terminology in this document may be selectively applicable to younger cohorts of children. Other influences impacting bladder function and continence include the developmental level of the child[18] as well as any behavioral disorders[15].

Storage symptoms

Increased or decreased voiding frequency

Voiding frequency is variable and is influenced by age[19] as well as by diuresis and fluid intake[20] more so than bladder capacity. At school-age children, 95% of children will void between 3 to 8 times per day[21] and population surveys document that most children will void between 3 to 7 times daily.[22, 23] Therefore we continue to propose the definition of increased daytime urinary frequency for children who void $\geq$ 8x per day and decreased daytime urinary frequency for children who void $\leq$ 3x per day. Voiding frequency may not be fully appreciated until a formal voiding frequency/volume chart or voiding diary is collected.

Incontinence

Incontinence (urinary incontinence) means involuntary leakage of urine and can be continuous or intermittent. The subdivisions of incontinence include continuous incontinence, intermittent incontinence, daytime incontinence and enuresis. (Figure 2).

Continuous incontinence refers to constant urine leakage and is most frequently associated with congenital malformations (i.e., ectopic ureter,

exstrophy variant) or functional loss of the external urethral sphincter function (e.g. external sphincterotomy) or to iatrogenic causes (e.g. vesicovaginal fistula). *Intermittent incontinence* is the leakage of urine in discrete amounts. Intermittent incontinence that occurs while awake is termed *daytime incontinence*. When intermittent incontinence occurs exclusively during sleeping periods, this is termed *enuresis*. Children with combined intermittent incontinence during awake periods and while sleeping would be termed: daytime incontinence and enuresis. For subdivisions of enuresis and daytime incontinence the reader is referred to the section on LUT conditions below.

Urgency

Urgency refers to the sudden and unexpected experience of an immediate and compelling need to void. The term is not applicable before the attainment of bladder control. The symptom of urgency is often a sign of detrusor overactivity.

Nocturia

Nocturia is the complaint that the child has to wake at night to void. Nocturia is common among school children[21, 24] and is not necessarily indicative of LUT dysfunction or a pathologic condition. Unlike enuresis, nocturia does not result in incontinence. Note that the term nocturia does not apply to children who wake up for reasons other than a need to void, for instance children who wake up after an enuretic episode.

Voiding Symptoms

Hesitancy

Hesitancy denotes difficulty in the initiation of voiding yet the child is ready to void.

Straining

Straining means that the child complains of needing to make an intensive effort to increase intra-abdominal pressure (e.g. Valsalva) in order to initiate and maintain voiding.

Weak Stream

This term is used to describe the observed stream or flow of urine with a weak force.

Intermittency

Intermittency is the term applied when micturition does not occur in a continuous stream but rather in several discrete stop and start spurts.

Dysuria

Dysuria is the complaint of burning or other discomfort during micturition. The timing of dysuria may be noted during voiding. Dysuria at the start of voiding

suggests a urethral source of pain and dysuria at the completion of the voiding suggests a bladder source of pain.

Other Symptoms

Holding Maneuvers

These are observable strategies used to postpone voiding or suppress urgency that may be associated with bladder overactivity. The child may or may not be fully aware of the purpose of the maneuvers, but they are usually obvious to caregivers. Common maneuvers include standing on tiptoes, forcefully crossing the legs, grabbing or pushing on the genitals or abdomen and placing pressure on the perineum (e.g. squatting with the heel pressed into the perineum or sitting on the edge of a chair).

Feeling of incomplete emptying

This term refers to the complaint that the bladder does not feel empty after voiding and may result in the need to return to the toilet to void again.

Urinary retention

This refers to the sensation of inability to void despite persistent effort.

Post micturition dribble

This term is used when the child describes involuntary leakage of urine immediately after voiding has finished. This symptom may be associated with vaginal reflux in girls or syringocoele in boys (see below).

Spraying (splitting) of the urinary stream

This term refers to the complaint that urine passage is a spray or a split rather than a single discrete stream. This usually implies a mechanical obstruction of encroachment of the meatus (e.g. meatal stenosis).

Genital and LUT pain

Bladder pain

Complaint of suprapubic pain or pressure or discomfort related to the bladder.

Urethral pain

Complaint of pain felt in the urethra

Genital pain

This term refers to pain in the genitals. In girls, vaginal pain and vaginal itching are commonly seen with localized irritation from incontinence. Penile pain and episodic priapism may be seen in young boys as symptoms associated with a full bladder or constipation.

TOOLS OF INVESTIGATION

A thorough history and physical examination are the hallmark diagnostic tools for evaluation of children and adolescents with LUT dysfunction. During the evaluation process, it is advisable to observe the child for any holding maneuvers, need to go to the toilet or any behavioral issues during the dynamics of the office visit. There are specific tools that aid the evaluation of pediatric LUT function and for a detailed description of these tools, please refer to the published ICCS guideline on the diagnostic evaluation of children with daytime incontinence.[13] We will briefly review these tools and their relevant terminology. These investigative tools will be categorized into invasive and non-invasive urodynamics.

Non-invasive urodynamics

Diaries

Bladder diary

The objective recording and documentation of bladder function involves collecting a diary. The full bladder diary consists of a 7-night recording of incontinence episodes and night-time urine volume measurements to evaluate enuresis, and a 48 hours daytime frequency and volume chart (which do not need to be recorded on 2 consecutive days) to evaluate for LUT dysfunction. Details of these documents can be found on the ICCS website and guidelines on evaluation for enuresis and LUT dysfunction.[8, 12, 13]

Bowel diary

The close relationship between bladder and bowel function requires screening of the bowel function to rule out BBD. The recommended work up for bowel dysfunction in the context of BBD is outlined in the ICCS guideline on the management of functional constipation in children with lower urinary tract symptoms.[9] Briefly, a 7-day bowel diary is advisable and includes the Bristol Stool Form Scale. The diagnosis of functional constipation in children is controversial and the Rome-III criteria is the most commonly accepted guideline for diagnosis.

Questionnaires

Questionnaires have emerged as a useful tool in the evaluation of LUT function. This need for questionnaires is largely based on the symptomatic nature of LUT dysfunction and the need to objectively translate subjective complaints into semi-quantitative data. The scoring of questionnaires allows care providers to gauge the impact of the LUT dysfunction and provides a method of monitoring outcomes during the treatment. Questionnaires are divided into 2 categories – LUT function questionnaires and Psychological screening questionnaires.

LUT function questionnaires

Several questionnaires have emerged as tools for the assessment of pediatric LUT function. Two LUT questionnaires stand out amongst these tools as they have been developed across cultures, validated and undergone test and re-testing for reliability.[25-29] These include:

Dysfunctional Voiding Symptom Score (DVSS)[25]: The DVSS questionnaire is utilized to quantify the severity of LUTS.

Pediatric Urinary Incontinence Quality of Life Score (PIN-Q)[28]: The PIN-Q questionnaire measures the emotional impact that urinary incontinence has on a child.

Both tools, the PIN-Q and DVSS, are complementary and provide a clinically appropriate picture of LUTS and impact on QOL.[30]

Psychological Screening

The high rate of comorbid clinical behavioral disorders associated with BBD is well documented and reviewed in detail in the ICCS document on psychological and psychiatric issues in urinary and fecal Incontinence.[15] The Child Behavior Checklist (CBCL) is a widely used parental questionnaire by psychiatrists and psychologists that contains 113 empirically derived behavioral items and has been translated into many languages. Any other validated, normed broad-band behavioral questionnaire can also be used (such as the Strengths and Difficulties Questionnaire (SDQ) of the Behavior Assessment for Children (BASC)).

Short Screening Instrument for Psychological Problems in Enuresis (SSIPPE) [31]

The SSIPE is a short screening instrument derived from the CBCL and recommended for the initial workup of a psychological problem associated with pediatric LUT dysfunction or BBD.

Urine Flow Measurement

Uroflow studies consist of measurement of urine flow with a uroflowmeter during voiding and are described in terms of rate and pattern, volume voided and post-void residual. To obtain an uroflow study, a child must be toilet trained before he or she can void into the flow meter. Additionally, it is important that there is an adequate volume of voided urine because uroflow curves change when voided volume is < 50% of expected bladder capacity for age.[13] Finally, it is important to obtain more than 1 curve to improve the accuracy of the uroflow tests and improve its interpretation.

Uroflowmetry may be done with or without electromyography (EMG) testing of the perineal muscles. The advantage of using EMG with uroflowmetry is the ability to appreciate synergy or dyssynergy between the bladder and the pelvic floor.

Flow rate

Maximum flow rate (Qmax) is the most relevant quantitative variable when assessing bladder outflow. Sharp peaks in the flow curve are usually artifacts, so maximum flow rate should be registered only at a peak level with a duration of at least two seconds.[32] In studies of normal children and adults, a linear correlation has been found between maximum flow and the square root of voided volume.[33] If the square of the maximum flow rate [$(ml/s)^2$] is equal to or exceeds the voided volume (ml), the recorded maximum flow is most probably within the normal range.

Flow curve shape

The shape of the flow curve is the most important factor to analyze during uroflowmetry when evaluating the flow curve of a child. The precise shape of the flow curve is determined by detrusor contractility, any abdominal straining and by the bladder outlet. Five major types of flow patterns are seen in children during uroflowmetry. (Figure 3). The flow pattern is not a guarantee to the underlying diagnostic abnormality but rather serves as a guide to the existence of a specific condition.

Bell-shaped curve.

The normal urinary flow curve of a healthy child is *bell-shaped* curve regardless of gender, age and voided volume.

Tower-shaped curve

This type of curve is a sudden, high-amplitude curve of short duration that suggests an overactive bladder producing an explosive voiding contraction.

Staccato-shaped curve

This flow pattern is irregular and fluctuating throughout voiding but the flow is continuous, never reaching zero during voiding. This flow pattern suggests sphincter overactivity during voiding and will be seen as sharp peaks and troughs in the flow curve. To qualify for the staccato label, the fluctuations should be larger than the square root of the maximum flow rate.

Interrupted-shaped curve

This flow curve will display discrete peaks with spikes similar to a staccato-shaped curve but unlike the staccato-flow pattern, there will be segments where zero flow with complete cessation between these peaks exist. This flow pattern suggests with an underactive bladder and each peak of flow represents an abdominal muscle strain creates the main force for bladder evacuation. In between the abdominal muscle straining, the flow will cease. It is also possible that this flow pattern can be seen with severe dyssynergy between the bladder and the external sphincter.

Plateau-shaped curve

This is a flattened, low-amplitude prolonged flow curve that is primarily

suggestive of a bladder outlet obstruction (BOO). The BOO can be anatomical (e.g. posterior urethral valves or urethral stricture) or dynamic (e.g. continuous, tonic sphincter contraction). Use of EMG during uroflowmetry may differentiate between BOO subtypes. A plateau-shaped curve may also be seen with an underactive bladder during a long continuous abdominal strain. Abdominal pressure monitoring during the uroflow can help delineate an underactive bladder condition.

Pelvic Ultrasound

The pelvic ultrasound is a key tool in the evaluation of pediatric LUT function[13] and terminology is applicable to the use of this investigative tool. Ultrasonography may be applicable as a bladder scan machine with a simple calculation of the post void residual (PVR) measurement or as a B-mode sonographic probe that can also provide anatomical details of the lower urinary tract as well as adjacent rectum.

Post void residual

Post void residual measurements in children are highly variable. Recent investigation of 1,128 healthy Taiwanese children between the ages of 4-12 years of age with a bell-shaped uroflow pattern and a voided volume of $\geq$ 50 ml support the following normative 95th percentile values for an abnormally elevated PVR[34]:

Children $\leq$ 6 years old: Single PVR > 30 ml or >21% bladder capacity (BC) where BC is expressed as the voided volume (VV) + PVR and expressed as percent of the expected bladder capacity (EBC = [age (yrs) + 1] x 30 ml)[1]. It is more optimal and recommended that a repeat PVR be performed and with dual PVR, a repetitive PVR > 20 ml or > 10% BC is considered significantly elevated.

Children $\geq$ 7 years old: A single PVR >20 ml or 15% BC, or repetitive PVR >10 ml or 6% BC is considered significantly elevated.

Standard parameters and conditions should be applied to measuring the PVR. These include that the bladder should not be under distended (< 50%) or over distended (>115%) in relation to the EBC. Additionally, PVR measurement should be obtained immediately after voiding (< 5minutes).

Bladder wall thickness

In daily clinical practice a thickened bladder wall alerts the clinician to longstanding problems with urine storage and emptying.[13] Bladder wall thickness can be measured with a full and empty bladder. However, normal values are not available. Bladder wall thickness depends on degree of bladder filling and it is likely that bladder wall thickness correlates with LUT dysfunction.[35]

Rectal distension

Similar to the bladder, there is insufficient evidence that the transverse

diameter of the rectum can be used solely as a predictor of constipation and fecal impaction.[9] In non-constipated and constipated children, a diameter greater than 30 mm correlated with a positive finding of rectal impaction on a digital rectal examination.[36]

Invasive urodynamics

Urodynamic studies are not routinely used to evaluate the LUT function in neurologically intact children[13] but are employed routinely in children suspected of having a neuropathic bladder[11]. A future ICCS document will detail pediatric urodynamic guidelines.

Urodynamic (cystometric) techniques

Urodynamic studies investigate filling and emptying phases of bladder function. Note that in the pediatric setting specific adaptations, regarding staff training, environment, child and parental support must be made to make the whole examination child-friendly. If the supra-pubic route is used for measuring bladder dynamics, a delay of time recommended between catheter insertion and urodynamic recording. If a transurethral catheter is used, the size of the catheter needs to be as small a diameter as possible to avoid outflow obstruction.

The term *cystometry* is used to describe the urodynamic investigation during the filling phase of the micturition cycle. The filling phase begins with the flow of fluid into the bladder and ceases when instillation ends. Several parameters of the filling phase should be identified in clinical reports and investigative studies. These defined cystometric parameters should state the filling rate, the temperature of the instilled fluid and the final volume of instilled fluid. The filling rate for pediatric studies should be close to physiologic filling and should approach approximately 5 to 10% of the EBC. The fluid temperature should be either room or body temperature. The total volume of fluid instilled should not exceed a volume that causes pain or results in prolonged passive detrusor pressures > 40 cm H2O.

Natural fill (ambulatory) cystometry provides the most physiologic simulation of bladder filling and the time and volumes should be identified during this urodynamic approach.

Bladder storage function should be described in terms of bladder sensation, detrusor activity, bladder compliance and bladder capacity.

Bladder sensation during filling cystometry

The determination of bladder sensation is subjective in young children and infants and less so in older children and adolescents. Physical cues (e.g. holding maneuvers) will be the clues for younger children who cannot express the sensation of a desire to void.

Reduced bladder sensation is defined as diminished sensation throughout bladder filling, and *absent bladder sensation* as no bladder sensation. Both conditions can be observed in children with an underactive detrusor, a neuropathic bladder or in co-morbid case of diabetes mellitus.

Detrusor function during filling cystometry

Normal detrusor function allows bladder filling with little or no change in pressure, and without involuntary detrusor contractions despite provocation such as coughing or positional changes. Thus, in infants and children any detrusor activity observed before voiding is considered pathological.

Detrusor overactivity is the occurrence of involuntary detrusor contractions during filling cystometry. These contractions may be spontaneous or provoked and produce a waveform on the cystometrogram of variable duration and amplitude. The contractions may be phasic or terminal. Symptomsof urgency and/or urgency incontinence may or may not occur. Similar to the latest IUGA/ICS terminology document[5], if a relevant neurological cause is present, then *neurogenic detrusor overactivity* is noted, otherwise *idiopathic detrusor overactivity* should be the preferred term.

Bladder capacity during filling cystometry

Cystometric capacity is the bladder volume at the end of filling cystometry, when ‘‘permission to void’’ is given during the urodynamics study. This endpoint and the level of the child’s bladder sensation at that time (‘‘normal desire to void’’) should be noted.

Maximum cystometric capacity is the bladder volume when the child can no longer delay micturition.

Bladder capacity during filling cystometry

Bladder compliance describes the relationship between a change in bladder volume and change in detrusor pressure. Compliance is calculated by dividing the volume change (ΔV) by the change in detrusor pressure ($\Delta Pdet$) during that change in bladder volume ($C = \Delta V/\Delta Pdet$).

Compliance is expressed as ml per cmH2O. Bladder compliance can be affected by several factors that should be standardized during the study including bladder filling and the reference points for compliance calculations. A faster filling rate is more provocative and as a general rule the filling rate (ml/min) should be between 5 and 10% of EBC and should not exceed 20 ml/min. The starting point for compliance calculations is the detrusor pressure at the start of bladder filling and the corresponding bladder volume (usually zero). The end point for compliance calculations is the passive detrusor pressure (and corresponding bladder volume) at cystometric capacity or immediately before the start of any detrusor contraction that causes significant leakage (and therefore causes the bladder volume to decrease).

In addition to the quantitative calculation, the shape of the filling curve is important and provides insight into bladder compliance. Normally, the detrusor pressures will remain relatively stable during bladder filling resulting in a linear-shaped curve. A non-linear shaped filling curve will be seen with rising detrusor

pressures during bladder filling. The change in the shape of the compliance curve should be noted at the corresponding bladder volume and time of the study as the change may occur early or later during bladder filling. For example, the overall quantitative compliance may be the same in two studies but one study has a nonlinear curve during the onset of filling but another study has a non-linear curve that occurs toward the end of bladder filling.

Urethral function during filling cystometry

Urethral function in children is usually assessed by pelvic floor EMG using skin or (less commonly) needle electrodes. The urethral closure pressure is rarely measured. For centers that use pressure measurements the IUGA/ICS definitions are applicable.[5] The occurrence of urethral leakage may differ when doing urodynamic studies in a supine position as compared to an upright position and thus body position should be noted.

Incompetent urethral closure mechanism is the leakage of urine that occurs during activities that raise intra-abdominal pressure in the absence of a detrusor contraction.

Urethral relaxation incontinence is defined as leakage due to urethral relaxation in the absence of raised abdominal pressure or detrusor contraction.

Urodynamic stress incontinence is the involuntary leakage of urine during filling cystometry, associated with increased intra-abdominal pressure (e.g. coughing or sneezing), in the absence of a detrusor contraction. In children, urodynamic stress incontinence is a less common condition as compared to adult females.

Leak point pressures

There are two types of leak point pressure measurement and the terminology for pediatrics is identical to the IUGA/ICS terminology[5]. The pressure values at leakage should be measured at the moment of leakage.

Detrusor leak point pressure (detrusor LPP): This static test is the lowest value of detrusor pressure at which leakage is observed in the absence of increased abdominal pressure or a detrusor contraction. High detrusor LPP (e.g.> 40 cmH2O) is associated with reduced bladder compliance and poses risk for upper urinary tract deterioration. High detrusor LPP is commonly denoted in children with a neuropathic bladder such as seen in spina bifida or other neurological disorders. There is no data on correlation between detrusor LPP and upper tract damage in children with a non-neuropathic bladder.

Abdominal leak point pressure (abdominal LPP): This is a dynamic test that measures the lowest value of intentionally increased intravesical pressure that provokes urinary leakage in the absence of a detrusor contraction. Coughing or Valsalva are examples of ways to induce increased pressure. A low abdominal LPP is suggestive of poor urethral function

Voiding cystometry (Pressure flow studies)

Voiding cystometry is the pressure-volume relationship of the bladder during micturition. Voiding cystometry can be evaluated in neurologically intact or near-intact infants and children but is less frequently performed due to its invasive nature and resultant distress.

Detrusor function during voiding

Normal detrusor function is characterized by an initial (voluntary) relaxation of the external urethral sphincter/pelvic floor followed immediately by a continuous detrusor contraction that leads to complete bladder emptying within a normal time span, and in the absence of obstruction.

Detrusor underactivity is voiding contraction of reduced strength and/or duration, resulting in prolonged bladder emptying and/or a failure to achieve complete emptying within a normal time span. An *acontractile detrusor* is seen when no contraction whatsoever occurs during urodynamic testing; the term *neurogenic acontractile detrusor* should be used where a neurological cause exists.

There are selective times when pressure-flow studies are of clinical value in children so as to distinguish between two clinical conditions that will result in low urine flow on uroflowmetry - an *underactive bladder vs*. *bladder outlet obstruction* (BOO). With an underactive bladder, there is detrusor underactivity where as with BOO, the detrusor pressure is elevated. An underactive bladder may involve abdominal straining to achieve complete micturition; consequently abdominal pressure may be elevated during micturition resulting in an interrupted uroflow curve.

Urethral Function During Voiding Cystometry

Normal urethral function: The urethra opens and is continuously relaxed to allow micturition at a normal pressure, urine flow, and no PVR.

Dysfunctional voiding is characterized by an intermittent and/or fluctuating flow rate due to intermittent contractions of the peri-urethral striated or levator ani muscles during voiding in neurologically normal children. A uroflow with electromyography (EMG) or a videourodynamic study is required to document dysfunctional voiding. The EMG is necessary to distinguish an interrupted or intermittent uroflow pattern secondary to an acontractile or underactive detrusor with abdominal voiding.

Detrusor sphincter dyssynergia (DSD) is incoordination between detrusor and sphincter during voiding (i.e., detrusor contraction synchronous with contraction of the urethral and/or periurethral striated muscles). This may be seen in either neurological voiding disorders and non-neuropathic LUT problems and is characterized by a “spinning-top” configuration of the proximal urethra on videocystourethrography (VCUG).

Four hour voiding observation

Four hour voiding observation is a validated technique used for the evaluation of bladder function in infancy. The method involves continuous observation of the freely moving infant with frequent ultrasound measurement of

bladder filling and residual urine after each voiding. Voided volumes may also be measured by the weighing of diapers.

Signs

Signs related to voided volumes

The term *voided volume* is used to characterized the volume of urine measured with micturition and is the volume recorded on the voiding record or diary. The voided volume is non-invasive and is a reflection of real life. It is of utmost importance because it is easy to obtain and determines followup treatment. Any other measure of bladder volume should contain an explanation of the method regarding how it was obtained e.g. ultrasound volume, urodynamic volume, catheterized volume, cystographic volume or cystoscopic volume.

The term *maximum voided volume* (MVV) refers to the largest volume of voided urine measured on the frequency volume chart but is variable if the first morning void is included. It is recommended that inclusion or exclusion of the first morning void be noted during investigations of the MVV. The term *expected bladder capacity* (EBC) is used as a reference or standard for comparison. The EBC is defined by the formula: (30 x [age in yrs + 1] ml).[37] This EBC formula was recently validated when the first morning void was disregarded on the frequency volume chart.[24] The EBC is applicable during 4 to 12 years of age as it reaches a level of 390 ml at 12 years. Finally, MVV, excluding the first morning void, is considered small or large if found to be < 65% or > 150% of EBC, respectively.

Signs related to urine output

Normal urine output is difficult to define in childhood, due to great intra- and inter-individual variation and to a lack of large-scale investigations. Following the lead of the IUGA/ICS, we use the term *polyuria* to describe excessive excretion of urine resulting in profuse and frequent micturition.[5] Polyuria will quantitatively be defined as voided urine volumes of $\geq$ 40 ml/kg body weight during 24 hr or $\geq$ 2.8 L urine for a child or adolescent weighing 70 kg or more.

Nocturnal urine output excludes the last voiding before sleep but includes the first voiding in the morning. In enuretic children, urine voided during sleep is collected in diapers and the change of diaper weight is measured. *Nocturnal polyuria* is a term relevant mainly in children suffering from enuresis and is defined in this cohort as a nocturnal urine output exceeding 130% of EBC for the child's age. We recognize there is a need for investigation to better determine the quantitative threshold of this definition. In a recent population-based study of 148 healthy children with 1,977 overnight recordings, nocturnal polyuria was found to be a urine volume > than 20 x (age + 9) in ml.[24] This latest formula may be applicable for a population based nocturnal polyuria, the clinical usefulness of which has not yet been tested. Accordingly, nocturnal polyuria will result in nocturia or enuresis. However, due to the necessary arbitrariness of this definition we strongly recommend authors studying these conditions report

nocturnal urine output and EBC, or the ratios between them, rather than merely classifying the children as polyuric or non-polyuric.

Conditions/Diagnosis

Using the ICD-10 and DSM-V definitions and criteria[7, 16], a condition of incontinence requires a minimum age of 5.0 years, a minimum of one episode per month and a minimum duration of 3 months to be termed a condition. Applying the criteria set forth by the DSM-5 and ICD-10, enuresis and daytime urinary incontinence is a significant condition if there is $\geq$ 1 episode per month and a frequency of 3 episodes over 3 months We further propose to qualify the significance of enuresis as frequent ($\geq$ 4 per week) or infrequent (<4 per week).

Enuresis

Enuresis is both a symptom and a condition of intermittent incontinence that occurs exclusively during sleeping periods. We propose to qualify the significance of enuresis as frequent ($\geq$ 4 per week) or infrequent (<4 per week).

Subgroups

There is an abundance of evidence that enuretic children with concomitant symptoms of LUT dysfunction differ clinically, therapeutically and pathogenetically from children without such daytime symptoms.[8, 12] Enuresis in children without any other LUT symptoms (nocturia excluded), and without bladder dysfunction, is defined as *monosymptomatic enuresis*. Other children with enuresis and any LUT symptoms are said to suffer from *non-monosymptomatic enuresis*. Subgrouping of enuresis into monosymptomatic and non-monosymptomatic is therefore essential and based on the current clinical situation. Once all daytime LUT symptoms have disappeared in an enuretic child, his/her enuresis switches from non-monosymptomatic to monosymptomatic.

If enuresis is subdivided according to its onset, the term *secondary enuresis* should be reserved for those children who have had a previous dry period of exceeding six months.[12] Otherwise the term *primary enuresis* should be used. A caveat for subtyping secondary enuresis is that it is associated with behavioral co-morbidities that necessitate investigation.

Daytime conditions

The classification of daytime LUT conditions is more complex than enuresis because of the heterogeneity of symptoms of LUT dysfunction and the considerable overlap between conditions. Additionally, borderline cases are common and the rationale for the grouping of various symptom complexes into specific LUT dysfunction conditions is often not fully evidence-based.

To provide a framework to classify daytime LUT dysfunction conditions, we recommend assessment and documentation be based on the following parameters:

1) Incontinence (presence or absence, and symptom frequency)
2) Voiding frequency
3) Voiding urgency

4) Voided volumes
5) Fluid intake

This is more important than subgrouping the children into various recognized conditions listed below. Although formally we use 5 years of age as the age of reference for symptoms and LUT conditions[7, 16], these conditions including incontinence are applicable to the age of attained bladder control.

Bladder and bowel dysfunction (BBD)

BBD is a condition of combined bladder and bowel disturbance that encompasses LUT and bowel function. *Severe BBD* is LUT and bowel dysfunction that is characteristic of the dysfunction seen in children with neurologic conditions yet has no identifiable or recognizable neurologic abnormality. This is synonymous with the historical term Hinman syndrome.

Overactive bladder

Overactive bladder (OAB): Urinary urgency, usually accompanied by frequency and nocturia, with or without urgency urinary incontinence, in the absence of urinary tract infection (UTI) or other obvious pathology. Children with OAB usually have *detrusor overactivity*, but this label cannot be applied to them without cystometric evaluation (see above). *Urgency incontinence* is the complaint of involuntary loss of urine associated with urgency and is thus a term that is applicable to many children with OAB.

Voiding postponement

Children who are observed by their parents and/or caregivers to habitually postpone micturition using holding maneuvers are said to suffer from *voiding postponement*. This behavior derived by clinical history is often associated with a low micturition frequency, a feeling of urgency and possibly incontinence due to a full bladder. Some children have learned to simultatneously restrict fluid intake so as to reduce incontinence. The rationale for delineating this entity lies in the observation that these children often suffer from psychological comorbidity or behavioral disturbances such as oppositional defiant disorder (ODD).[15]

Underactive bladder

This is a clinical term that is reserved for children with the need to raise intra-abdominal pressure to initiate, maintain or complete voiding, i. e. straining. The children may have low voiding frequency in the setting of adequate hydration but may also have frequency due to incomplete emptying with prompt refilling of the bladder. These children often produce an interrupted uroflow pattern and are usually found to have *detrusor underactivity* if examined with invasive urodynamics. Flow patterns may also be plateau-shaped; pressure–flow studies will distinguish it from bladder outlet obstruction.

Dysfunctional voiding

The child with *dysfunctional voiding* habitually contracts the urethral sphincter or pelvic floor during voiding and demonstrates a staccato pattern with

or without an interrupted flow on repeated uroflow when concomitantly recording EMG activity.

Bladder outlet obstruction (BOO)

BOO is a condition that refers to the impediment flow of urine during voiding. BOO may be mechanical or functional, static or phasic. BOO is characterized by increased detrusor pressure and a reduced urine flow rate during pressure-flow studies.

Stress incontinence

Stress incontinence is the involuntary leakage of small amounts of urine with effort or physical exertion that increases intra-abdominal pressure e.g. coughing or sneezing. During urodynamic investigation, the leakage is confirmed in the absence of a detrusor contraction and termed *urodynamic stress incontinence*.

Vaginal reflux

Toilet-trained girls who consistently experience daytime incontinence in moderate amounts shortly after normal voiding and have no underlying mechanism other than obvious vaginal entrapment of urine are said to have *vaginal reflux*. This is not associated with other LUT symptoms or enuresis. It is a consequence of voiding with legs closed leading to urine entrapment inside the introitus. It may be seen in girls with labial adhesions.

Giggle incontinence

Giggle incontinence is a rare condition in which complete voiding occurs specifically during or immediately after laughing. Bladder function is normal when the child is not laughing. Giggle incontinence is not linked to any other disturbance of LUT function.

Extraordinary daytime urinary frequency

This term applies to toilet-trained children who have a sudden onset of voiding often associated with small volumes during the daytime only. The daytime voiding frequency is at least once per hour and average voided volumes are less than 50% of EBC (usually much smaller). Incontinence is not often associated with this condition and nocturia is absent. Co-morbidities such as polydipsia, diabetes mellitus UTI or viral syndrome should be excluded.

Bladder neck dysfunction

Bladder neck dysfunction refers to the condition where there is an impaired/delayed opening of the bladder neck resulting in impaired flow despite an adequate or elevated detrusor contraction.[38] The prolonged opening time, i.e., the time between the start of a voiding detrusor contraction and the start of urine flow can be seen on videourodynamics. Alternatively bladder neck dysfunction

can be diagnosed non-invasively with a uroflow/EMG when a prolonged EMG lag time, i.e., the time interval between the start of pelvic floor relaxation and the actual start of flow.[38] The EMG lag time remains to be further defined and validated.

Comorbidity

It is not the task of the ICCS to suggest definitions and terminology for areas outside the LUT. We do, however, find it useful to list comorbid conditions that are relevant and important to take into account for researchers studying the LUT in children. These include the following:

- Constipation and fecal incontinence
- Urinary tract infection
- "Asymptomatic" bacteriuria
- Vesicoureteral reflux
- Neuropsychiatric conditions (ADHD, oppositional defiant disorder etc.)
- Intellectual disabilities
- Disorders of sleep (sleep apneas, parasomnias)
- Body mass index

Of special relevance are behavioral disorders, which affect 20-40% of children with enuresis and 30-40% with day-time incontinence. These include externalizing disorders such as ADHD and ODD, as well as internalizing disorders such as depressive and anxiety disorders.[15]

Treatment

Definitions of treatment methods

ICCS treatment guidelines have been published in documents defining various LUT conditions and comorbidities.[8-10, 12, 14, 15] This document conveys definitions and guidelines regarding terminology alone.

We strongly advise writers not to use terms such as "standard therapy" or "maintenance therapy" without defining the design of these treatments.

Pharmacological therapy, surgical therapy

These terms mean any therapy based on drugs or surgery.

Neuromodulation

This term refers to therapy that reduces LUT symptoms or restores LUT function by the alteration and modulation of nerve activity through central and/or peripheral electrical stimulation or chemical agents to targeted sites.

Alarm treatment

Alarm treatment is therapy based on a device that gives a strong sensory signal – usually, but not necessarily, acoustic – immediately after an incontinence episode. It can be used during day- or nighttime, although the latter usage is more common.

Urotherapy

Urotherapy is conservative-based therapy and treatment of LUT dysfunction. It is LUT rehabilitation and encompasses a very wide field of health-care professionals. Urotherapy can be divided into standard therapy and specific interventions.

Urotherapy initially encompasses the following components:

1) *Information and demystification*. Explanation about normal LUT function and in what way the particular child deviates from normal
2) *Instruction* in what to do about LUT dysfunction; i.e. behavioral modification with regular voiding habits, proper voiding posture, avoidance of holding maneuvers, etc.
3) *Life-style advice*. Advice regarding fluid intake, proper diet (prevention of constipation), etc.
4) *Registration* of symptoms and voiding habits, using bladder diaries or frequency-volume charts
5) *Support* and *encouragement* via regular follow-up with the caregiver

Specific interventions of urotherapy are defined similarly to that published by the ICS[4], and includes various forms of pelvic floor muscle training (which includes biofeedback), neuromodulation and intermittent catheterization. Additional specific interventions of urotherapy involve cognitive behavioral therapy (CBT) and psychotherapy.

Psychotherapy encompasses all non-surgical, non-pharmacological treatments aimed at comorbid behavioral and emotional disorders accompanying incontinence (but not aimed at enuresis or urinary incontinence themselves). These evidence-based techniques are indicated following thorough child psychological or psychiatric assessment and only if a behavioral disorder is present. They can be augmented by pharmacotherapy (such as stimulants in ADHD). The treatment of these comorbid emotional and behavioral disorders does not only alleviate suffering for the child and his/her family, but can increase compliance and adherence to urotherapy – leading to improved outcomes.[15]

Definitions of treatment outcome

In the scenario, the affected child and family are the ones who decide appropriate criteria for treatment success. In the research setting, however, a uniform standard is necessary, so that studies and treatment options can be compared.

Three basic principles of treatment outcomes should be recognized by researchers:

1) The symptom frequency during baseline and following treatment should each be documented.
2) The assessment of treatment response or outcome must be based on pre-treatment baseline registration of the frequency of symptoms.
3) The response during treatment should be noted as well as the response after cessation of treatment for a specified period of time. These responses may not be the same.

Initial success

No-response: <50% reduction
Partial response: 50 to 99% reduction.
Complete response: 100% reduction

Long-term success

Relapse: more than one symptom recurrence per month
Continued success: no relapse in 6 months after interruption of treatment
Complete success: no relapse in two years after interruption of treatment

LEGENDS

Figure 1. Bladder and bowel dysfunction subtypes.

Figure 2. Incontinence subtypes.

Figure 3. Uroflow curve patterns. A. Bell-shaped pattern. B. Tower-shaped pattern. C. Staccato-shaped pattern. D. Interrupted-shaped pattern. E. Plateau-shaped pattern.

FIGURES

Figure 1.

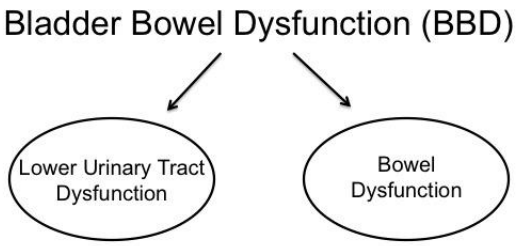

Figure 2.

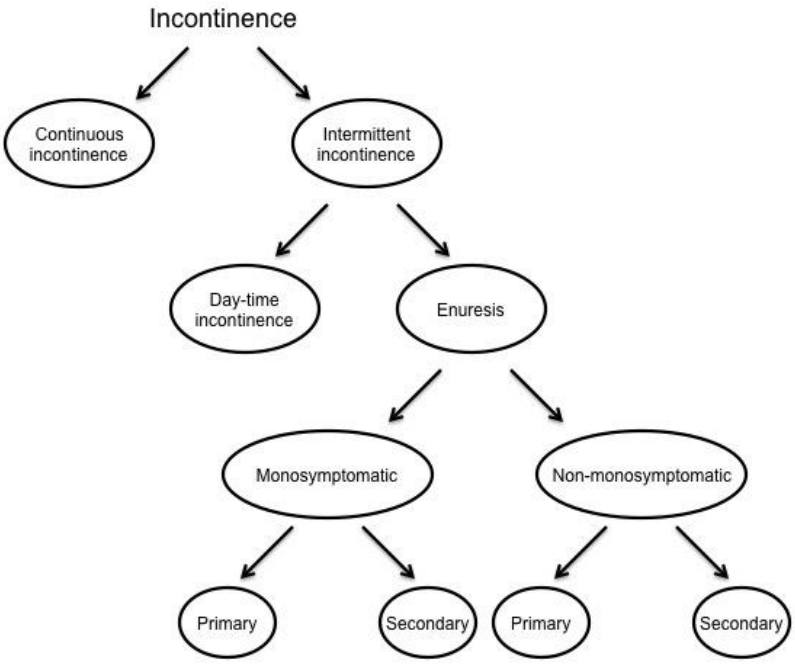

Figure 3.

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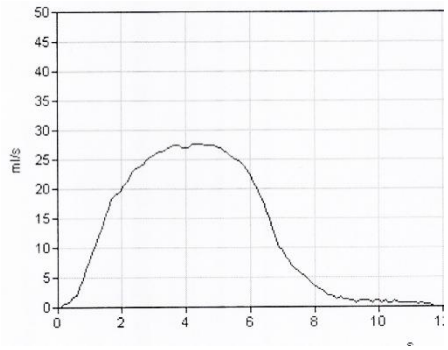
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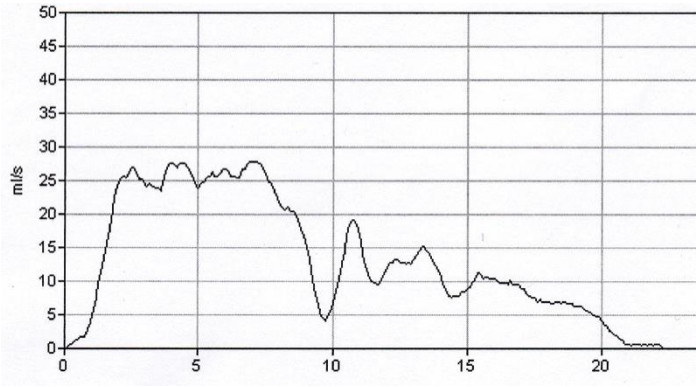
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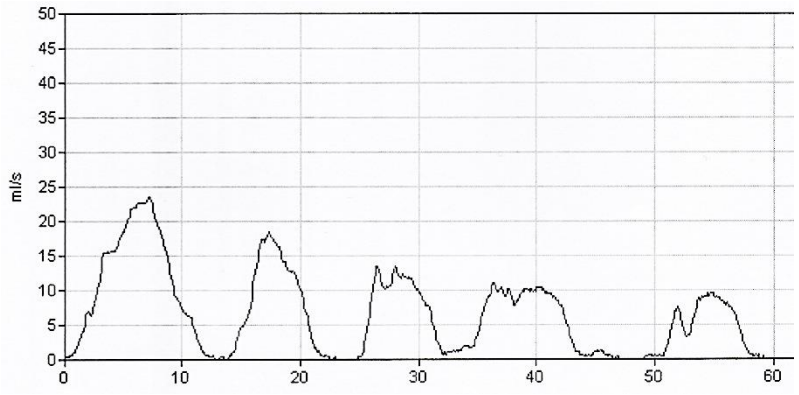
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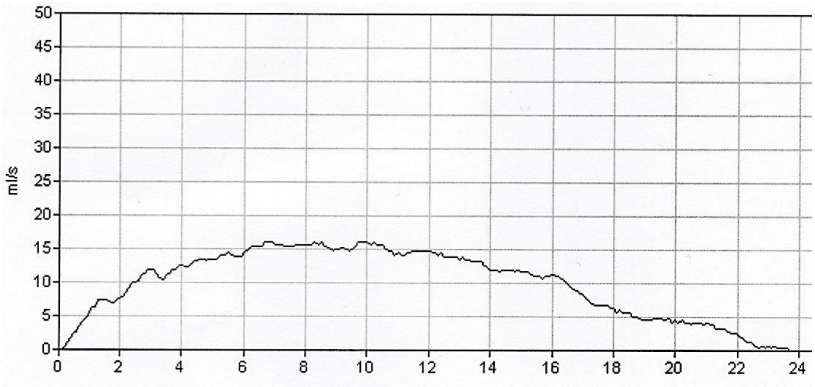
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