Neurourology and Urodynamics

The Standardization of Terminology of Lower Urinary Tract Function in Children and Adolescents: Update Report From the Standardization Committee of the International Children's Continence Society

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Aim: The impact of 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 across multiple healthcare disciplines. The present document serves as a stand-alone terminology update reflecting refinement and current advancement of knowledge on pediatric LUT function. **Methods:** A variety of worldwide experts from multiple disciplines *within the ICCS leadership* who care for children with LUT dysfunction were assembled *as part of the standardization committee*. A critical review of the previous ICCS terminology document and the current literature was performed. Additionally, contributions and feedback from the multidisciplinary ICCS membership were solicited. **Results:** Following a review of the literature over the last 7 years, the ICCS experts assembled a new terminology document reflecting 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. *Neurourol. Urodynam. 9999:1–11,* 2015.

Key words: child; consensus; terminology; urinary bladder/physiology; urination

INTRODUCTION

The standardization of terminology for pediatric bladder and bowel function is critical in providing a platform for optimal understanding, communication and treatment across multiple health care providers who care for children and adolescents with lower urinary tract (LUT) dysfunction. Terminology that is applicable internationally is particularly pertinent due to 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 terminology of pediatric LUT dysfunction is thus imperative to reduce confusion among providers. Standardized terms are also critical for comparing research and study outcomes to optimally promote investigative understanding of pediatric LUT dysfunction.

The ICCS is a unique organization whose members comprise multiple disciplines and specialties from almost every continent that care for children with bladder and bowel incontinence. Thus, the ICCS is uniquely positioned to provide guidance in the standardization of terminology for bladder and bowel dysfunction (BBD) in children and adolescents.

Over the last decade, the second report from the Standardization Committee of the ICCS[1] has propagated definitions and established standardized terminology that allowed for clarity

Abbreviations: ADHD, attention deficit hyperactivity disorder; BBD, bladder bowel dysfunction; BC, bladder capacity; BOO, bladder outlet obstruction; CBCL, Child Behavior Checklist (CBCL); DSD, detrusor sphincter dysfunction; DSM-5, Fifth Edition of the Diagnostic and Statistical Manual of Mental Disorders; DVSS, Dysfunctional Voiding Symptom Score; EBC, expected bladder capacity; EMG, electromyography; ICCS, International Childrens Continence Society; ICD-10, International Classification of Diseases-10; ICS, International Continence Society; IUGA, International Urogynecological Association; LPP, leak point pressure; LUT, lower urinary tract; MVV, maximum voided volume; OAB, overactive bladder; PVR, post void residual.

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of communication. 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) that focus on pediatric LUT function. 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 adopting of ICCS terminology. Despite this significant impact of the global usage of ICCS terminology, approximately 25% of studies published between 2007 and 2010 contained 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 ICCS terminology for pediatric bladder and bowel function serves as a stand-alone terminology update reflecting refinement and advancement of knowledge on these systems. Adherence to the updated terminology is followed at all ICCS courses and workshops and it is encouraged that all investigators and clinicians who publish on this topic utilize the ICCS recommended terminology. To delineate manuscripts and publications that follow the ICCS guidelines regarding terminology we recommend future manuscripts include the text "Terminology adheres to standards recommended by the ICCS except where specifically noted."

MATERIALS AND METHODS

A variety of worldwide experts from multiple disciplines who care for children with LUT dysfunction were assembled. The standardization committee consisted of active members and leaders of the ICCS that have extensively published on several facets of BBD and all of the ICCS documents published in the last 4 years. Healthcare disciplines included urology, nephrology, gastroenterology, general and developmental pediatrics, physical therapy, psychology, and psychiatry. The standardization committee emanated from North and South America, Europe, the Middle East, Africa, Australia, and Asia. A critical review of the original ICCS terminology document and the current literature was performed. Additionally, input from the multidisciplinary ICCS membership was solicited.

This terminology document represents the 3rd published standardization on terminology for LUT function and enhances previous ICCS documents.[1,3] Recognition and reference to the terminology on LUT 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 to be current and inclusive of other global organizations and disciplines that also deal with continence. Additionally, terms and definitions employed by the new Fifth Edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5)[6,7] were considered and the ICD-10 medical classification list from the World Health Organization[8] was referenced.

This update is not intended to serve as a guideline for clinical treatment. There are numerous previous ICCS documents outlining treatment for specific LUT and associated co-morbid conditions.[9–16] This terminology update follows the prior ICCS terminology outline of establishing syntax to properly convey symptoms of LUT dysfunction and to affirm terminology for investigative tools, signs, conditions, and treatment parameters as they pertain to LUT function and dysfunction. The reader is referred to the prior ICCS communications for a comprehensive description of the pathophysiology. 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 whose primary expertise is in urinary continence and bladder function but equally acknowledge a close relationship between bowel and bladder function. Thus, the importance of bowel related terms in relation to bladder function is emphasized.

TERMINOLOGY

Bladder and Bowel Dysfunction (BBD)

Due to the aforementioned relationship between the bladder and bowel, concomitant bladder and bowel disturbances have been labeled as BBD. We discourage using the term dysfunctional elimination syndrome (DES) as this connotes a particular abnormality or condition. We recommend BBD as a more descriptive comprehensive term of a combined bladder and bowel disturbance that does not explain pathogenesis but rather encompasses this parallel dysfunction. BBD is an umbrella term that can be subcategorized into LUT dysfunction and bowel dysfunction (Fig. 1).

When the term dysfunction or disorder is used, it represents clinical significance and relevance. In a research document or reference, authors should specify and provide support for using the term BBD. In the absence of any co-morbid bowel dysfunction, the term LUT dysfunction alone suffices.

Symptomatic Terms

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 and duration of a symptom may alter the perception of its relevance. Nevertheless, duration of time is beneficial in characterizing symptoms.

Terminology used for LUT symptoms will focus on descriptive rather than quantitative language, as quantitative data to define symptomatic terms is lacking. Age of the child is particularly relevant when applying terminology for pediatric bladder function. Our reference point for LUT symptoms is >5 years of age as this age is used by the DSM-5 and the International Classification of Diseases-10 (ICD-10) to characterize urinary incontinence disorders.[6,8] For functional bowel dysfunction the minimum age is 4 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; therefore, this terminology document may be selectively applicable to younger cohorts of

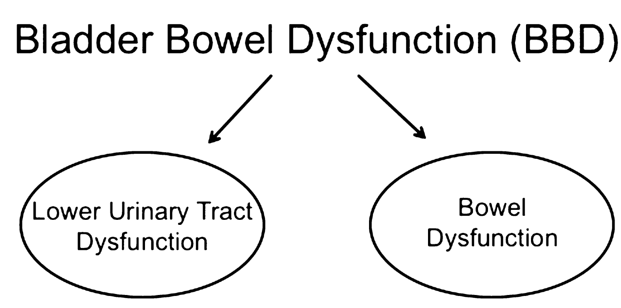

Fig. 1. Bladder and bowel dysfunction subtypes.

children. Other influences impacting bladder function and continence include the developmental level of the child[18] as well as any behavioral disorders.[12]

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. Normative data in population surveys are mixed. In a small, cross-sectional analysis of healthy school-aged children, approximately 95% of 7–15 years old children will void between 3 to 8 times per day[21]; population surveys in larger sample sizes report that most 7 year olds will void between 3 to 7 times daily[22] whereas in another large population survey most children between 3–12 years of age void 5–6 times per day.[23] Based on the large surveys and the previous terminology document,[1] the panel continues to propose the definition of increased daytime urinary frequency in those children who void $\geq 8 \times$ per day and decreased daytime urinary frequency for the ones who void $\leq 3 \times$ per day. Voiding frequency may not be fully appreciated unless a formal voiding frequency/volume chart or voiding diary is collected.

Incontinence. Urinary incontinence means involuntary leakage of urine; it can be continuous or intermittent. The subdivisions of incontinence include continuous incontinence, intermittent incontinence, daytime incontinence and enuresis. (Fig. 2).

Continuous incontinence refers to constant urine leakage (day and nighttime) usually associated with congenital malformations (i.e., ectopic ureter, exstrophy variant), functional loss of the external urethral sphincter function (e.g., external sphincterotomy) or 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, it is termed *enuresis*. Enuresis should not be used to refer to daytime incontinence. A child with combined intermittent incontinence during "awake" periods and while sleeping is termed daytime incontinence and enuresis. For subdivisions of enuresis and daytime incontinence, the reader is referred to the sections on Conditions/Diagnosis (Enuresis) and LUT symptoms 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 bladder 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 nocturia does not apply to children who wake up for reasons other than a need to void, for example, children who wake up after an enuretic episode.

Voiding Symptoms

Hesitancy. Hesitancy denotes difficulty in initiating voiding when the child is ready to void.

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

Weak stream. This term describes an observed stream or uroflow that is weak.

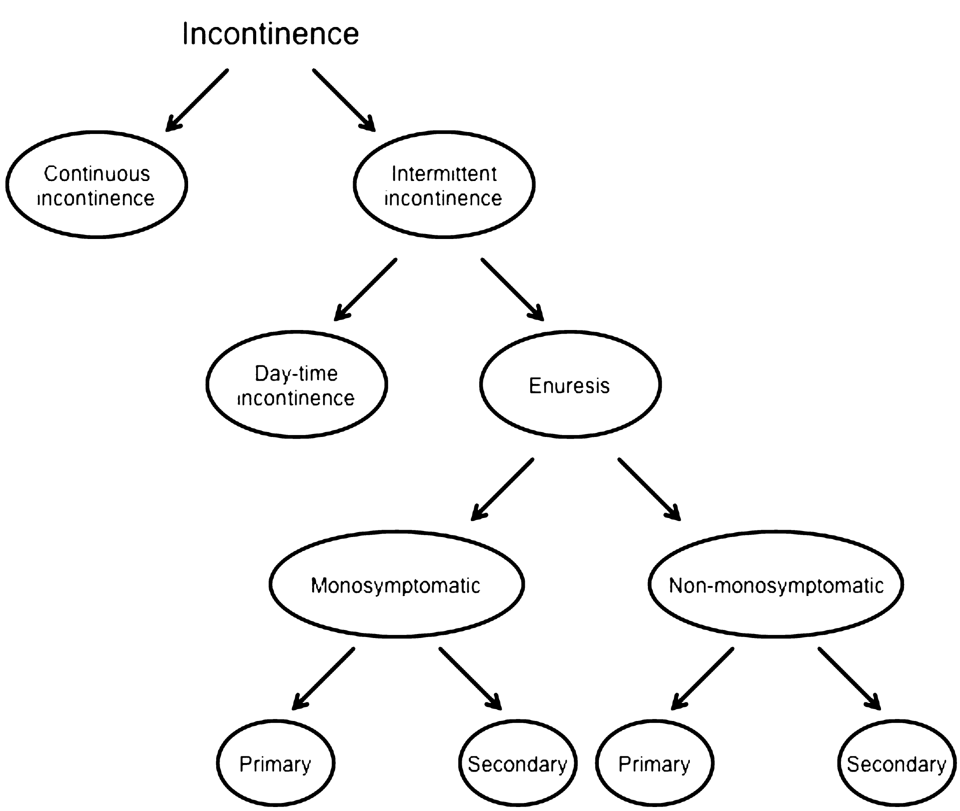

Fig. 2. Incontinence subtypes.

Intermittency. Intermittency implies micturition that is not continuous but rather has several discrete stop and start spurts.

Dysuria. Dysuria is the complaint of burning or discomfort during micturition. The timing of dysuria may be noted during voiding. Dysuria at the start of voiding suggests a urethral source of pain whereas dysuria at the completion of voiding suggests a bladder.

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 these maneuvers, but they are usually obvious to caregivers. Common behaviors 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 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 an inability to void despite persistent effort in the presence of a fully, distended bladder. Duration of time is particularly beneficial in characterizing retention.

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 refers to the complaint that urine passes as a spray or a split rather than a single discrete stream. It usually implies a mechanical obstruction at or just below 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 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, constipation or the result of urine trapping inside a phimotic foreskin.

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, it is advisable to observe the child for holding maneuvers, expressions of urgency or any behavioral issues. Specific tools that aid the evaluation have been published in the ICCS guideline on diagnostic evaluation of children with daytime incontinence.[10] These tools and their relevant terminology will be briefly reviewed and 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. A *complete bladder diary* consists of a 7-night recording of incontinence episodes and nighttime urine volume measurements to evaluate enuresis, and a *48 hr daytime frequency and volume chart* (not necessarily recorded on two consecutive days) to evaluate for LUT dysfunction. Details can be found on the ICCS website (http://www.i-c-c-s.org) and guidelines on evaluation for enuresis and LUT dysfunction.[10,11,16] Mobile device applications (apps) may also facilitate bladder diary recordings.

Bowel diary. The close relationship between bladder and bowel function requires screening of both systems to rule out BBD. The 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 LUT symptoms.[15] A 7-day bowel diary utilizing the Bristol Stool Form Scale is preferable. The diagnosis of functional constipation in children is controversial; the Rome-III criteria are the most commonly accepted guideline for diagnosis.

Questionnaires

Questionnaires have emerged as useful adjuncts in the evaluation of LUT function. This need is largely based on the symptomatic nature of LUT dysfunction and the importance of objectively translating subjective complaints into semi-quantitative data. The scoring of questionnaires allows providers to gauge the extent of the dysfunction and provides a method of monitoring outcomes during treatment. Two types of questionnaires exist—measurements of LUT function and psychological screening.

LUT Function Questionnaires

Although several questionnaires have emerged as assessment tools, two stand out as they have been tested across cultures, validated and undergone test and re-testing for reliability.[25–29] These include:

Dysfunctional voiding symptom score (DVSS).[25] The DVSS questionnaire quantifies severity of LUTS.

Pediatric urinary incontinence quality of life score (PIN-Q).[28] The PIN-Q measures the emotional impact that urinary incontinence has on a child.

Both tools are complementary and provide a clinically appropriate picture of LUTS and impact on quality of life.[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.[12] The Child Behavior Checklist (CBCL) is a widely used parental questionnaire by psychiatrists and psychologists that contains 113 empirically derived behavioral items. The CBCL has been translated into several languages. Any validated, normed broadband behavioral questionnaire can be used that is, 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 brief instrument derived from the CBCL and recommended initially if any psychological problem associated with pediatric LUT dysfunction or BBD exists.

Urine Flow Measurement

Uroflow studies consist of measuring the rate, volume voided, voiding time and examining the pattern during urination into an uroflowmeter. To obtain an uroflow, a child must obviously be toilet trained. Additionally, it is important (1) the volume of voided urine is adequate as curves change when voided volume is <50% of expected bladder capacity for age[10] and (2) to obtain more than one curve to improve accuracy, reliability and interpretation of the test.

Uroflowmetry may be done with or without electromyography (EMG) testing of the perineal muscles. The advantage of combining 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 curve are usually artifacts, so maximum flow rate should be registered only when a peak level has a duration of >2 sec.[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 [$(ml/s)^2$] equals or exceeds the voided volume (ml), the recorded maximum flow is most probably normal.

Flow curve shape. The shape of the flow curve is paramount when analyzing the flow pattern. The precise shape is determined by detrusor contractility and influenced by abdominal straining, coordination with the bladder outlet musculature and any distal anatomic obstruction. Five types of flow patterns are seen. (Fig. 3). Each specific pattern is no guarantee of an underlying diagnostic abnormality but rather serves as a guide to the existence of a specific condition.

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

Tower-shaped curve. This is a sudden, high-amplitude curve of short duration that suggests an overactive bladder produced by 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 pattern suggests incoordination of the bladder and the sphincter with intermittent sphincter overactivity during voiding (i.e., dysfunctional voiding). It will be seen as sharp peaks and troughs in the flow curve. To qualify for a staccato label, the fluctuations should be larger than the square root of the maximum flow rate.

Interrupted-shaped curve. This flow will display discrete peaks with spikes similar to a staccato-shaped curve but unlike the latter pattern, there will be segments where zero flow with complete cessation between these peaks exists. This flow pattern suggests an underactive bladder; each peak represents abdominal muscle straining creating the main force for urine evacuation. In between each strain, the flow ceases. It is possible this flow pattern can be seen with incoordination between the bladder and external urethral sphincter.

Plateau-shaped curve. This is a flattened, low-amplitude prolonged flow curve that is suggestive of 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). Flow electromyography (EMG) may differentiate between BOO subtypes. A plateau-shaped curve may 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

Pelvic ultrasound is a key tool in the evaluation of pediatric LUT function.[10] Ultrasonographic bladder scan machines calculates bladder volume, and thus are useful in measuring pre- and post void residual (PVR) or as a B-mode sonographic probe that provides anatomical details of the LUT and adjacent rectum.

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

Children 4–6 years old. Single PVR >30 ml or >21% of bladder capacity (BC) where BC is determined as voided volume (VV) + PVR and expressed as percent of the expected bladder capacity (EBC = [age (yrs) + 1] × 30 ml)[1]. It is recommended that a repeat PVR be performed with dual measurements, a repetitive PVR >20 ml or >10% BC is considered significantly elevated.

Children 7–12 years old. A single PVR >20 ml or 15% BC, or repetitive PVR >10 ml or 6% BC is considered elevated.

Standard conditions should be applied to measuring PVR: the bladder should not be under-distended (<50%) nor over-distended (>115%) in relation to the EBC; PVR must be obtained immediately after voiding (<5 min). Further validation is needed for the above nomograms in similar cohorts across cultures.

Bladder Wall Thickness

In daily clinical practice a thickened bladder wall alerts the clinician to longstanding problems with urine storage and emptying.[10] Bladder wall thickness can be measured with a full and empty bladder. However, normal values do not exist. Bladder wall thickness depends on degree of bladder filling. It is likely that bladder wall thickness correlates with LUT dysfunction.[35]

Rectal Distension

There is insufficient evidence that the transverse diameter of the rectum can be used solely as a predictor of constipation and fecal impaction.[15] In non-constipated and constipated children, a diameter >30 mm on pelvic ultrasound correlated with a finding of rectal impaction on a digital rectal examination.[36]

Invasive Urodynamics

Urodynamic studies are not routinely used to evaluate LUT function in neurologically intact children[10] but are employed regularly in children suspected of having a neuropathic bladder[13]. A future ICCS document will detail pediatric urodynamic guidelines.

Urodynamic (cystometric) techniques. Urodynamic studies investigate filling and emptying phases of bladder function. In the pediatric setting, there should be specific adaptations regarding staff training, environment, child, and parental support so the entire examination is child–friendly. If bladder dynamics are measured via a suprapubic catheter, a delay of time is recommended between catheter insertion and urodynamic recording. If a transurethral catheter is used, catheter size needs to be as small as possible to avoid outflow obstruction.

Cystometry is used to describe the urodynamic investigation during the filling phase of the micturition cycle. Before filling is started, the bladder must be emptied completely. The filling phase begins with the flow of fluid into the bladder and ceases

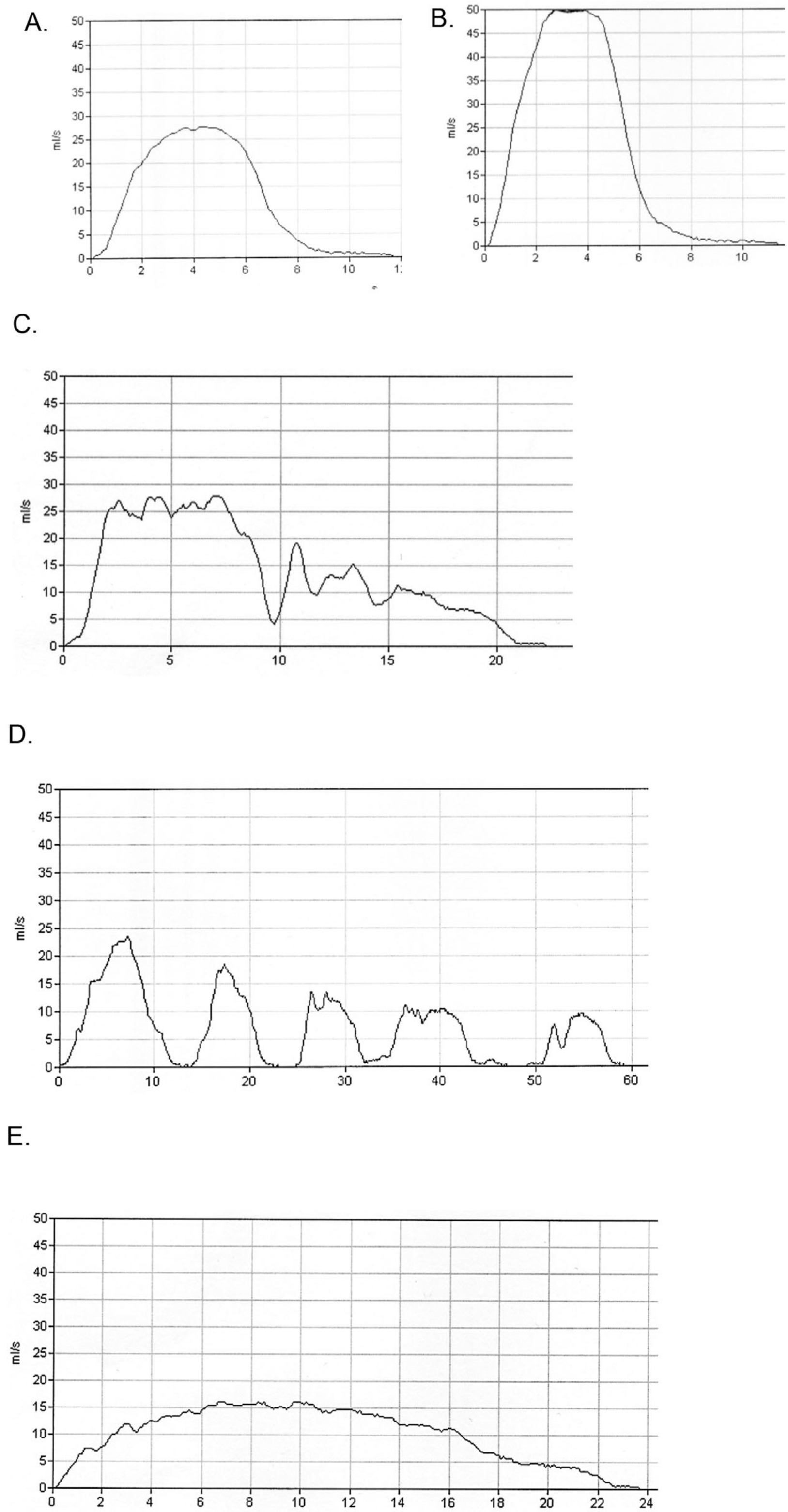

Fig. 3. Uroflow curve patterns. (A) Bell-shaped. (B) Tower-shaped. (C) Staccato-shaped. (D) Interrupted-shaped. (E) Plateau shaped.

when instillation ends. Several parameters during this phase should be identified in the clinical report that includes the filling rate, temperature of the infusate and the final volume instilled. The filling rate should be close to physiologic filling approaching 5–10% of EBC. Fluid temperature should be between 25 and 37 °C and the volume of instilled fluid should not exceed an amount that causes pain or results in prolonged passive detrusor pressures >40 cm H_2O.

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

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

Bladder sensation during filling cystometry. Bladder sensation is subjective in infants and toddlers but less so in older children and adolescents. Physical cues (e.g., holding maneuvers) will be the signs in younger children who cannot express the sensation or a desire to void.

Reduced bladder sensation is defined as diminished awareness throughout bladder filling, and *absent bladder sensation* as no bladder sensation whatsoever. Both can be observed in children with an underactive detrusor, a neuropathic bladder or a co-morbidity 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. 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. They may be spontaneous or provoked and produce a waveform of variable duration and amplitude. Contractions may be phasic or terminal. Symptoms of urgency and/or urgency incontinence may or may not occur. Similar to the latest IUGA/ICS terminology,[5] if a relevant neurological cause is present, then *neurogenic detrusor overactivity* is noted, otherwise *idiopathic detrusor overactivity* is 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 childs bladder sensation at that time ("normal desire to void") should be noted.

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

Bladder compliance during filling cystometry. *Bladder compliance* describes the relationship between changes in bladder volume and changes 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 cm H_2O. Bladder compliance can be affected by several factors that should be standardized during the study such as the rate of filling and the reference points for compliance calculations. A faster filling rate is more provocative and should not exceed 5–10% of EBC or 20 ml/min. The starting point for compliance calculations is the detrusor pressure at the initiation 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 (that causes the bladder volume to decrease).

In addition to the quantitative calculation, the shape of the filling curve is important; it provides insight into bladder compliance. Normally, detrusor pressure remains relatively stable throughout bladder filling resulting in a linear shaped curve. A non-linear shaped filling curve will be seen with rising detrusor pressure during filling. The change in shape of the compliance curve should be noted at the corresponding bladder volume and time of the study as change may occur early or later during bladder filling. The overall quantitative compliance may be similar in two studies but one study has a nonlinear curve during the onset of filling whereas another has a nonlinear curve that occurs toward the end of bladder filling.

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

Incompetent urethral closure mechanism is leakage of urine occurring 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; the terminology for pediatrics is identical to IUGA/ICS terminology.[5] The pressure values at leakage should be measured at the moment it occurs.

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 muscle compliance and poses risk for upper urinary tract deterioration. High detrusor LPP is commonly denoted in children with a neuropathic bladder, that is, spina bifida or related neurological disorders. It should be noted that if a patient has little to no bladder neck or intrinsic sphincter function, then the DLPP is not an accurate reflection of detrusor compliance. Subsequently, bladder wall compliance is further assessed with maneuvers to increase the outlet resistance. 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 inducing increased pressure. A low abdominal LPP is suggestive of poor urethral function. Abdominal LPP supplants the terms Valsalva or stress LPP.

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, in the absence of obstruction.

Detrusor underactivity denotes a 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 in order to distinguish between two clinical conditions that will result in low flow on uroflowmetry—an *underactive bladder versus BOO*. With the former, there is detrusor underactivity whereas with BOO, the detrusor pressure is elevated. An underactive bladder may require abdominal straining to achieve complete micturition; consequently abdominal pressure may be elevated during voiding 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 and flow with 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. An uroflow with 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 external urethral sphincter muscles during voiding (i.e., detrusor contraction synchronous with contraction of the urethral and/or periurethral striated muscles). This is seen in neurological disorders on urodynamic evaluation and is characterized by increased EMG sphincter activity during a detrusor contraction and by either a "spinning-top" configuration of the proximal urethra or a narrowing of the external sphincter area on videocystourethrography (VCUG) or videourodynamics.

A "spinning-top" urethra may also be seen in neurologically intact children with incoordination of the external sphincter and bladder during voiding (i.e., dysfunctional voiding) on VCUG. Additionally, it should be noted that patients with OAB without dysfunctional voiding might exhibit a "spinning-top" urethral appearance due to habitual guarding or holding maneuvers during increased bladder pressure or urgency.

Four Hour Voiding Observation

Four hour voiding observation is a validated technique used to evaluate bladder function during infancy.[1] This involves continuous observation of the freely moving infant with frequent ultrasound measurement of bladder filling and residual urine before and after each void. Voided volumes may also be calculated by weighing of diapers.

Signs

Signs related to voided volumes. The term *voided volume* is used to characterize the volume of urine measured with micturition and is recorded on the voiding diary. Voided volume is non-invasive and reflective of real life. It is of utmost importance because it is easy to obtain and influences follow-up treatment. Any other measure of bladder volume should explain the method used to obtain it, for example, ultrasound, urodynamic, catheterized, cystographic, or cystoscopic volume.

The term *maximum voided volume* (MVV) refers to the largest volume of voided urine measured on the frequency volume chart throughout a 24-hr cycle. It is variable if the first morning void is included. It is recommended that inclusion or exclusion of the first morning void be noted during investigation 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: ($30 \times$ [age in yrs $+ 1$] ml).[37,38] This EBC formula was recently validated when the first morning void was disregarded on the frequency volume chart.[24] The EBC is applicable for children between 4 and 12 years 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. As the IUGA/ICS noted, the term *polyuria* is used to describe excessive excretion of urine resulting in profuse and frequent micturition.[5] Polyuria is defined as voided urine volumes of >40 ml/kg body weight during 24 hr or >2.8 L urine for a child or adolescent weighing >70 kg .

Nocturnal urine output excludes the last voiding before sleep but includes the first morning void. In enuretic children, urine voided during sleep is collected in diapers and the change in diaper weight is measured. *Nocturnal polyuria* is relevant in children suffering from enuresis and is defined in this cohort as a nocturnal urine output exceeding 130% of EBC for age. There is a need to investigate 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 when urine volume was greater than $20 \times (\text{age} + 9)$ in ml.[24] This latest formula may be applicable for a population based nocturnal polyuria, but its clinical usefulness has yet to be tested. Accordingly, nocturnal polyuria will result in nocturia or enuresis. However due to the necessary arbitrariness of this definition, it is recommended for authors studying these conditions to 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,[6,8] the symptom of incontinence requires a minimum age of 5 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 it occurs >1 episode per month and a frequency of three episodes over 3 months We further propose to qualify the significance of enuresis as frequent (>4 per week) or infrequent (<4 per week).

Enuresis. Enuresis is both a symptom and a condition of intermittent incontinence that occurs during periods of sleep.

Subgroups

There is ample evidence that enuretic children with concomitant symptoms of LUT dysfunction differ clinically, therapeutically and pathogenically from children without such daytime symptoms.[11,16] Enuresis without other LUT symptoms (nocturia excluded), and without bladder dysfunction, is defined as *monosymptomatic enuresis*. Children with enuresis and any LUT symptoms are said to have *non-monosymptomatic enuresis*. Subgrouping of enuresis in this manner is essential and based on the *current* clinical situation. In patients with non-monosymptomatic enuresis, the type of LUT dysfunction condition should be reported, because this information will influence the treatment and the reproducibility of the data. Once daytime LUT symptoms have abated, the enuresis switches from non-monosymptomatic to monosymptomatic.

If enuresis is subdivided according to its onset, *secondary enuresis* is reserved for those children who have had a previous dry period of >6 months.[11] Otherwise it is termed *primary enuresis*. A caveat for subtyping secondary enuresis is its association with behavioral co-morbidities that necessitate investigation.

Daytime Conditions

The classification of daytime LUT conditions is more complex than enuresis due to the heterogeneity of symptoms of LUT dysfunction and the considerable overlap between conditions. Borderline cases are common; the rationale for grouping various symptom complexes into specific LUT dysfunction is often not adequately evidence-based.

To provide a framework to classify daytime LUT dysfunction, assessment and documentation should 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 the age of reference for symptoms and LUT conditions is >5 years,[6,8] these conditions including incontinence are applicable to the age of attained bladder control.

Bladder and bowel dysfunction (BBD). BBD is a combination of bladder and bowel disturbances. *Severe BBD* is characterized by LUT and bowel dysfunction seen in children with neurologic conditions who have no identifiable or recognizable neurologic abnormality. When severe BBD results in changes in the upper urinary tract (e.g., hydronephrosis and/or vesicoureteral reflux), it may be synonymous with the historical term "Hinman syndrome."

Overactive bladder. *Overactive bladder* (OAB): Urinary urgency, usually accompanied by frequency and nocturia, with or without urinary incontinence, in the absence of urinary tract infection (UTI) or other obvious pathology. Children with OAB usually have *detrusor overactivity*, but this label can only be applied with cystometric evaluation (see above). *Urgency incontinence* is the complaint of involuntary loss of urine associated with urgency and is thus applicable to many children with OAB.

Voiding postponement. Children who habitually postpone micturition using holding maneuvers 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 from a full bladder. Some children learn to simultaneously restrict fluids so as to reduce their 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).[12]

Underactive bladder. This clinical term is reserved for children who need to raise intra-abdominal pressure to initiate, maintain or complete voiding that is, 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 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 repeat uroflow when EMG activity is concomitantly recorded. Note: This is a term associated with a neurologically intact patient.

Bladder outlet obstruction (BOO). BOO refers to an impediment of urine flow during voiding. It may be mechanical or functional, static or phasic and is characterized by increased detrusor pressure and a reduced urinary 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, for example, coughing or sneezing. During urodynamic investigation, 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 voiding and have no other LUT symptoms or nighttime incontinence have *vaginal reflux*. It is a consequence of voiding with adducted legs leading to urine entrapment inside the introitus. It may be associated with labial adhesions due to localized inflammation.

Giggle incontinence. Giggle incontinence is a rare condition in which extensive emptying or leakage occurs during or immediately after laughing. Bladder function is normal when there is no laughter.

Extraordinary daytime only urinary frequency. This applies to a toilet-trained child who has the frequent need to void that is associated with small micturition volumes solely during the day. The daytime voiding frequency is at least once per hour with an average voided volume of <50% of EBC (typically 10–15%). Incontinence is rare and nocturia is absent. Co-morbidities, that is, polydipsia, diabetes mellitus, nephrogenic diabetes insipidus, daytime polyuria, UTI or viral syndrome, should be excluded.

Bladder neck dysfunction. *Bladder neck dysfunction* refers to impaired/delayed opening of the bladder neck resulting in reduced flow despite an adequate or elevated detrusor contraction.[39] The prolonged opening time, that is, the time between the start of a detrusor voiding contraction and the start of urination can be seen and measured on videourodynamics. Alternatively bladder neck dysfunction can be diagnosed non-invasively with a uroflow/EMG when a prolonged EMG lag time is noted, that is, the time interval between the beginning of pelvic floor relaxation and the actual start of flow.[39] 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 beyond the LUT. We do, however, find it useful to list comorbid conditions that are relevant and important, especially for researchers studying the LUT in children. These include the following:

- Constipation and fecal incontinence
- Urinary tract infection

- "Asymptomatic" bacteriuria
- Vesicoureteral reflux
- Neuropsychiatric conditions (attention deficit hyperactivity disorder (ADHD), oppositional defiant disorder etc.)
- Intellectual disabilities
- Disorders of sleep (sleep apneas, parasomnias)
- Obesity

Of special relevance are behavioral disorders, which affect 20–40% of children with enuresis and 30–40% with daytime incontinence. These include externalizing disorders (ADHD and ODD), and internalizing disorders (depressive and anxiety disorders).[12]

TREATMENT

Definitions of Treatment Methods

ICCS treatment guidelines have been published in documents defining various LUT conditions and comorbidities.[9,11,12,14–16] This document conveys definitions and guidelines regarding terminology alone.

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

Pharmacological therapy, surgical therapy. These pertain to any therapy based on drugs or surgery.

Neuromodulation. This 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 night-time, although the latter usage is more common.

Urotherapy. Urotherapy is conservative-based therapy and treatment of LUT dysfunction that rehabilitates the LUT and encompasses a very wide field of healthcare professionals. Urotherapy can be divided into standard therapy and specific interventions.

Urotherapy encompasses the following standard components:

(1) *Information and demystification*. Explanation about normal LUT function and how the particular child deviates from normal.

(2) *Instruction* in how to resolve LUT dysfunction; that is, behavioral modification with regular voiding habits, proper voiding posture, avoidance of holding maneuvers, regular bowel habits, etc.

(3) *Life-style advice*. Encompasses balanced fluid intake and diet, diminished caffeine, regular bladder and bowel emptying patterns, etc.

(4) *Registration* of symptoms and voiding habits, using bladder diaries or frequency–volume charts and potentially mobile apps.

(5) *Support* and *encouragement* via regular follow-up with the caregiver.

Specific interventions of urotherapy are defined similar to ICS guidelines[4] that include various forms of pelvic floor muscle retraining (biofeedback), neuromodulation, and intermittent catheterization. Additional 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 psychological or psychiatric assessment and only if a behavioral disorder is present. They can be augmented by pharmacotherapy (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.[12]

Definitions of treatment outcome

In the clinical 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.

Researchers should recognize three basic principles of treatment outcomes:

(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 pretreatment 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.

Note: The term "Response" (>90% reduction) has been dropped and rolled into the term "Partial response" to simplify and strengthen the term "Complete response."

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.

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