



Complete Summary

GUIDELINE TITLE

Obstructive pathology of renal duplication: ureterocele and ectopic ureter. In: Guidelines on paediatric urology.

BIBLIOGRAPHIC SOURCE(S)

Obstructive pathology of renal duplication: ureterocele and ectopic ureter. In: Tekgul S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R, Radmayr C, Stein R. Guidelines on paediatric urology. Arnhem, The Netherlands: European Association of Urology, European Society for Paediatric Urology; 2008 Mar. p. 63-7. [25 references]

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

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METHODOLOGY - including Rating Scheme and Cost Analysis
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SCOPE

DISEASE/CONDITION(S)

Obstructive pathology of renal duplication:

- Ureterocele
- Ectopic ureter

GUIDELINE CATEGORY

Diagnosis
Evaluation

Management
Treatment

CLINICAL SPECIALTY

Pediatrics
Surgery
Urology

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

- To outline a practical and preliminary approach to paediatric urological problems
- To increase the quality of care for children with urological problems

TARGET POPULATION

Infants and children with obstructive pathology or renal duplication (ureterocele and ectopic ureter)

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis/Evaluation

1. Classification of ureterocele according to degree of obstruction and functional impairment
2. Diagnosis of ureterocele
 - Prenatal ultrasound with confirmatory ultrasound at birth
 - Clinical symptoms, beside incidental findings
 - Ultrasonography
 - Nuclear renography
 - Magnetic resonance (MR) urography
 - Voiding cystourethrography (VCUG)
 - Urethrocystoscopy
3. Diagnosis of ectopic ureter
 - Clinical symptoms
 - Ultrasonography, nuclear studies, VCUG, MR urography
 - Methylene blue filling of the bladder

Treatment/Management

1. Treatment of ureterocele
 - Endoscopic decompression
 - Partial nephro-ureterectomy
 - Complete primary reconstruction
 - Follow-up, with secondary surgery as necessary

- Prophylactic antibiotic in cases of early diagnosis in clinically asymptomatic patients
2. Treatment of ectopic ureter
 - Heminephro-ureterectomy
 - Ureteral reconstruction

MAJOR OUTCOMES CONSIDERED

Successful decompression or reconstruction

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

The guidelines were based on current literature following a systematic review using MEDLINE.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

1a Evidence obtained from meta-analysis of randomized trials

1b Evidence obtained from at least one randomized trial

2a Evidence obtained from at least one well-designed controlled study without randomization

2b Evidence obtained from at least one other type of well-designed quasi-experimental study

3 Evidence obtained from well-designed non-experimental studies, such as comparative studies, correlation studies and case reports

4 Evidence obtained from expert committee reports or opinions or clinical experience of respected authorities

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Application of a structured analysis of the literature was not possible due to a lack of well-designed studies. Whenever possible, statements have been classified in terms of level of evidence and grade of recommendation. Due to the limited availability of large randomized controlled trials – influenced also by the fact that a considerable number of treatment options relate to surgical interventions on a large spectrum of different congenital problems – this document is therefore largely a consensus document.

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

- The first step in the European Association of Urology (EAU) guidelines procedure is to define the main topic.
- The second step is to establish a working group. The working groups comprise about 4-8 members, from several countries. Most of the working group members are academic urologists with a special interest in the topic. In general, general practitioners or patient representatives are not part of the working groups. A chairman leads each group. A collaborative working group consisting of members representing the European Society for Paediatric Urology (ESPU) and the EAU has gathered in an effort to produce the current update of the paediatric urology guidelines.
- The third step is to collect and evaluate the underlying evidence from the published literature.
- The fourth step is to structure and present the information. The strength of the recommendation is clearly marked in three grades (A-C), depending on the evidence source upon which the recommendation is based. Every possible effort is made to make the linkage between the level of evidence and grade of recommendation as transparent as possible.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Grades of Recommendation

- A. Based on clinical studies of good quality and consistency addressing the specific recommendations and including at least one randomized trial
- B. Based on well-conducted clinical studies, but without randomized clinical studies
- C. Made despite the absence of directly applicable clinical studies of good quality

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

There is no formal external review prior to publication.

The Appraisal of Guidelines for Research and Evaluation (AGREE) instrument was used to analyse and assess a range of specific attributes contributing to the validity of a specific clinical guideline.

The AGREE instrument, to be used by two to four appraisers, was developed by the AGREE collaboration (www.agreecollaboration.org) using referenced sources for the evaluation of specific guidelines. (See the "Availability of Companion Documents" field for further methodology information).

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Background

Ureterocele and ectopic ureter are the two main anomalies associated with a complete renal duplication. At present, antenatal ultrasonography detects both conditions in the majority of cases and diagnosis is confirmed after birth by further examination. Later on in life, these anomalies are revealed by clinical symptoms: urinary tract infection (UTI), pain, disturbances of micturition and urinary incontinence.

Ureterocele

Ureterocele is a cystic dilatation developed on the intravesical part of the ureter corresponding to the upper pole of a renal duplication. It is more frequent in females than in males, the overall prevalence being 1 in 4,000 births.

Ectopic Ureter

Ectopic ureter is less frequent than ureterocele (1 in 40,000 births), but is again more common in females (male to female ratio, 1:5). Eighty percent of ectopic ureters are associated with a complete renal duplication.

In females, the ureteral orifice may be located:

- In the urethra, from bladder neck to meatus (35%)
- In the vaginal vestibule (30%)
- In the vagina (25%)

- In the uterus and Fallopian tube (rarely)

In males, the ureteral orifice may be located:

- In the posterior urethra above the verumontanum and never below the external sphincter (60%)
- In the seminal tract (vas deferens, ejaculatory ducts, seminal vesicles) (40%)

Classification

Ureteroceleles are usually obstructive for the upper pole moiety, but the degree of obstruction and functional impairment is variable according to the type of ureterocele and upper pole dysplasia. In the orthotopic form, there is often no, or only mild, obstruction and frequently the function of the moiety is normal or slightly impaired; the corresponding ureter may be dilated. In the ectopic form, the upper pole is altered, frequently dysplastic and hypofunctional or non-functional. The corresponding ureter is a megaureter. In the caecoureterocele (see definition below), the upper pole of the renal duplication is always dysplastic and nonfunctional.

Ectopic Ureterocele

Ectopic ureterocele is the most common form of ureterocele (>80%) and occurs bilaterally in about 40% of cases. It is voluminous, dissociating the trigone and slipping into the urethra, and can rarely prolapse through the urethral meatus. The ureterocele orifice is tight, rarely large, and located near the bladder neck, either in the bladder itself or in the urethra below the bladder neck. The ureter corresponding to the lower pole moiety is raised by the ureterocele and is frequently refluxing or compressed by the ureterocele, leading to an obstructive megaureter. A contralateral renal duplication is associated in 50% of cases. Occasionally, very large ureteroceleles are responsible for reflux or obstruction of the contralateral upper tract.

Orthotopic Ureterocele

Orthotopic ureterocele accounts for 15% of cases. It is exclusively observed in females, and is small and located strictly intravesically. Orthotopic ureteroceleles are mostly combined with a single kidney system.

Caecoureterocele

Caecoureterocele is very rare, occurring in less than 5% of cases. It is associated with an ectopic ureter and located in the urethra below the bladder neck.

Diagnosis

Ureterocele

Prenatal ultrasound reveals easily voluminous obstructive ureteroceleles. In cases of a very small upper pole or a slightly obstructive ureterocele, prenatal diagnosis will be difficult. If prenatal diagnosis has been impossible, the following clinical

symptoms - beside incidental findings - can reveal the congenital anomaly at birth or later:

- At birth, a prolapsed and sometimes strangulated ureterocele may be observed in front of the urethral orifice. In a newborn boy, it might cause acute urinary retention, simulating urethral valves.
- The early symptom of pyelonephritis in either sex may lead to the diagnosis.
- Later symptoms can include dysuria, recurrent cystitis and urgency.

In cases of prenatal diagnosis at birth, ultrasonography confirms the ureteral dilatation ending at the upper pole of a renal duplication. It also demonstrates the presence of an ureterocele in the bladder, with a dilated ureter behind the bladder.

At this point, it is important to assess the function of the upper pole using nuclear renography in the region of interest. Magnetic resonance urography may visualize the morphological status of the upper pole and lower moieties and of the contralateral kidney. A voiding cystourethrography (VCUG) is mandatory in identifying an ipsilateral or contralateral reflux and to assess the degree of intraurethral prolapse of the ureterocele.

Urethrocystoscopy may reveal the pathology in cases where it is difficult to make the differential diagnosis between ureterocele and ectopic megaureter.

Ectopic Ureter

Most of the ectopic megaureters are diagnosed primarily by ultrasound. In some cases, clinical symptoms can lead to diagnosis:

- In neonates: dribbling of urine, pyuria and acute pyelonephritis
- An ectopic orifice may be found in the meatal region. Significant vaginal discharge may be the equivalent of incontinence in little girls
- In pre-adolescent males: epididymitis is the usual clinical presentation and a palpable seminal vesicle may be found on digital rectal examination

Ultrasonography, nuclear studies, voiding cystourethrogram (VCUG), magnetic resonance (MR) urography and cystoscopy are the diagnostic tools to assess function, to detect reflux and to rule out ipsilateral compression of the lower pole and urethral obstruction. In the clarification of incontinence in girls, a methylene blue filling of the bladder can be helpful: in the case of loss of uncoloured urine, an ectopic ureter must be present.

Treatment

Ureterocele

The management is controversial with the choice between endoscopic decompression, partial nephroureterectomy or complete primary reconstruction. The choice of a therapeutic modality depends on the following criteria: clinical status of the patient (e.g., urosepsis), age of the patient, renal function of the

upper pole, presence or absence of reflux, obstruction of the ipsilateral ureter, pathology of the contralateral ureter, and parents' and surgeon's preferences.

Early Diagnosis

- In a clinically asymptomatic child with an ureterocele and non- or hypo-functional upper pole, without significant obstruction of the lower pole and without bladder outlet obstruction, prophylactic antibiotic treatment is given until follow-up procedures are instigated
- In the presence of severe obstruction and infection, an immediate endoscopic incision or puncture is recommended

Re-evaluation

If decompression is effective and there is no reflux (approximately 25% of cases), the patient is followed-up conservatively. Secondary surgery is necessary if decompression is not effective, significant reflux is present or there is obstruction of the ipsi- or contra-lateral ureters and/or bladder neck obstruction. Surgery may vary from partial nephrectomy to complete unilateral reconstruction.

Ectopic Ureter

In the majority of cases, the upper pole is dysplastic and heminephro-ureterectomy should be considered. Ureteral reconstruction is a therapeutic option in cases in which the upper pole has function worth preserving.

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated for each recommendation.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate diagnosis, treatment, and management of ureterocele and ectopic ureter

POTENTIAL HARMS

Not stated

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

The purpose of these texts is not to be proscriptive in the way a clinician should treat a patient but rather to provide access to the best contemporaneous consensus view on the most appropriate management currently available. European Association of Urology (EAU) guidelines are not meant to be legal documents but are produced with the ultimate aim to help urologists with their day-to-day practice.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

The European Association of Urology (EAU) Guidelines long version (containing all 19 guidelines) is reprinted annually in one book. Each text is dated. This means that if the latest edition of the book is read, one will know that this is the most updated version available. The same text is also made available on a CD (with hyperlinks to PubMed for most references) and posted on the EAU websites Uroweb and Urosource (www.uroweb.org/professional-resources/guidelines/ & <http://www.urosource.com/diseases/>).

Condensed pocket versions, containing mainly flow-charts and summaries, are also printed annually. All these publications are distributed free of charge to all (more than 10,000) members of the Association. Abridged versions of the guidelines are published in European Urology as original papers. Furthermore, many important websites list links to the relevant EAU guidelines sections on the association websites and all, or individual, guidelines have been translated to some 15 languages.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Obstructive pathology of renal duplication: ureterocele and ectopic ureter. In: Tekgul S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R, Radmayr C, Stein R. Guidelines on paediatric urology. Arnhem, The Netherlands: European

Association of Urology, European Society for Paediatric Urology; 2008 Mar. p. 63-7. [25 references]

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2008 Mar

GUIDELINE DEVELOPER(S)

European Association of Urology - Medical Specialty Society
European Society for Paediatric Urology - Medical Specialty Society

SOURCE(S) OF FUNDING

European Association of Urology

GUIDELINE COMMITTEE

Not stated

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

All members of the working group submit a conflict of interest form. The information is kept on file in the European Association of Urology (EAU) Central Office database. This guidelines document was developed with the financial support of the EAU. No external sources of funding and support have been involved. The EAU is a non-profit organisation and funding is limited to administrative assistance, travel, and meeting expenses. No honoraria or other reimbursements have been provided.

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the [European Association of Urology Web site](#).

Print copies: Available from the European Association of Urology, PO Box 30016, NL-6803, AA ARNHEM, The Netherlands.

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- EAU guidelines office template. Arnhem, The Netherlands: European Association of Urology (EAU); 2007. 4 p.
- The European Association of Urology (EAU) guidelines methodology: a critical evaluation. Arnhem, The Netherlands: European Association of Urology (EAU); 18 p.

Print copies: Available from the European Association of Urology, PO Box 30016, NL-6803, AA ARNHEM, The Netherlands.

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI Institute on November 18, 2008. The information was verified by the guideline developer on December 19, 2008.

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