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ORIGINAL ARTICLE

Long-term renal outcome in infants with congenital lower urinary tract obstruction

Pronostic rénal à long terme des enfants présentant une obstruction sous-vésicale congénitale

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KEYWORDS
Lower urinary tract obstruction (LUTO);
Congenital anomalies of the kidney and the urinary tract (CAKUT);
Posterior urethral valves;
Paediatric urology;
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Summary
Introduction. — Congenital Lower Urinary Tract Obstructions (LUTO) is a heterogeneous group of diseases in which urine elimination is obstructed at the level of bladder neck or urethra. The aim of the study is to evaluate the long-term renal outcome of patients suffering of congenital LUTO.
Patients and method. — We retrospectively reviewed patients with congenital LUTO. All patients had at least 1 year follow-up. Data on surgery, renal imaging and Schwartz estimate creatinine clearance were collected. Incidence of Chronic Renal Disease (CRD) is presented with Kaplan-Meier method.
Results. — 40 patients were included, 23 patients with Posterior Urethral Valve (PUVs) and 17 patients with other aetiologies: anterior urethral valve (2), urethral atresia (2), urethral stenosis (2), cloacal malformations (2), obstructive ureterocele (1), bladder trigone malformation (1) and neonatal bladder-sphincter dysfunction without neurological abnormalities (7). Incidence of CRD at age 10 years was 37% in congenital LUTO, 42% in PUVs and 30% in other aetiologies, and was significantly higher in PUVs ($P = 0.032$). Renal prognosis was significantly worsened by discover of retentional bladder wall changes in initial cystoscopy, and by loss of parenchymal differentiation or cortical microcysts in first ultrasonography. The use of urinary diversion was significantly higher in LUTO of other aetiologies.

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Conclusion A high incidence of CRD is observed in patients with congenital LUTO, significantly higher in patients with PUV. LUTO of other aetiologies require step by step surgical management and higher use of urinary diversion. Precise initial evaluation in cystoscopy and ultrasonography is required and participate to evaluate future renal outcome.
Level of incidence. — 4.

MOTS CLÉS
Obstruction sous-vésicale congénitale (LUTO) ;
Anomalies congénitales du rein et des voies urinaires (CAKUT) ;
Valve de l'urètre postérieur ;
Urologie pédiatrique ;
Néphrologie pédiatrique

Résumé
Introduction. — Les obstructions sous-vésicales congénitales (OSVC) sont responsables d'un obstacle à l'élimination des urines au niveau du col vésical ou de l'urètre. Le but de l'étude est l'évaluation du pronostic rénal à long terme des patients présentant une OSVC.
Patients et méthode. — Étude rétrospective de patients présentant une OSVC avec un suivi post-natal minimal de 1 an. Les données chirurgicales, l'imagerie rénale et la clairance estimée de la créatinine selon Schwartz ont été collectées. L'incidence d'insuffisance rénale chronique (IRC) est présentée par une méthode de Kaplan-Meier.
Résultats. — Quarante patients ont été inclus, 23 présentant une valve de l'urètre postérieur (VUP) et 17 présentant un autre diagnostic: valve de l'urètre antérieur (2), atrésie uréthrale (2), sténose uréthrale (2), malformation cloacale (2), urétrocèle obstructive (1), malformation du trigone vésical (1) et dysfonction sphinctérienne vésicale non neurologique (7). L'incidence d'IRC était de 37 % à l'âge de 10 ans, significativement plus élevé chez les VUP (42 %) que chez les autres (30 %) ($p = 0,032$) Le pronostic rénal futur est assombri par la découverte d'une vessie de lutte ou d'une saillie du col vésical à la cystoscopie initiale, et par la dédifférenciation du parenchyme rénal ou la présence de microkystes corticaux à la première échographie.
Conclusion. — Une incidence élevée d'IRC est observée chez les patients présentant une OSVC, significativement plus élevée chez les patients avec VUP. Les autres étiologies présentent des anomalies associées nécessitant plus fréquemment chirurgies itératives et dérivation urinaire. L'évaluation initiale en cystoscopie et échographie doit être précise et participe à l'évaluation du pronostic rénal.
Niveau de preuve.— 4.

Introduction

Congenital Lower Urinary Tract Obstructions (LUTO) is a heterogeneous group of diseases in which urine elimination is obstructed at the level of bladder neck or urethra. This obstruction *in utero* leads to a global injury of the urinary tract. The mechanisms of such injuries are well known, associating dilatation and fibrosis of bladder and ureter and bilateral renal hypoplasia or dysplasia [1—4]. Congenital LUTO are the most severe group of Congenital Anomalies of the Kidney and the Urinary Tract (CAKUT). CAKUT are the first cause of pediatric Chronic Renal Disease (CRD) and End Stage Renal Disease (ESRD) [5—8].

Aetiologies of congenital LUTO are numerous and heterogeneous. The prevalence of LUTO is reported as 2.24 live births per 10,000 live births [9]. The most common etiology is Posterior Urethral Valves (PUVs), affecting only male fetuses and representing 63% of congenital LUTO. Other aetiologies of LUTO are urethral atresia, urethral stenosis, megalourethra, anterior urethral valves, Prune-Belly Syndrome, cloacal anomalies, obstructive ureterocele and severe congenital bladder-sphincter dysfunction [9].

Diagnosis of LUTO must be confirmed in emergency. The treatment is based on urinary diversion and/or obstacle ablation. Bladder catheterization, suprapubic stent, vesicostomy, ureterostomy or nephrostomy are options [10]. In PUVs, primary endoscopic valve ablation in the first days of life without bladder catheterization is the gold standard, permitting to better preserve bladder function [11]. Anaesthetic risk and surgical trauma must be considered in fragile patients for whom bladder catheterization becomes an option.

Long-term outcome of patients suffering of congenital LUTO is marked by renal impairment who determines the prognosis. In patients with congenital LUTO treated with Vesico Amniotic Shunt (VAS), the ESRD rate varies from 33% to 36% and the CRD rate is 22% [12,13]. In patients with PUVs, the incidence of ESRD is 10% at age 10 years and 38% at age 20 years, with highest incidence occurring in the first year of life and in late adolescence. The incidence of chronic renal failure is 34% at age 10 years, and 51% at age 20 years [11]. A review showed an ESRD rate of 11% (0—20%) and a CRD rate of 22% (0—32%) in patients with PUVs treated by endoscopic valve resection. In this study, elevated nadir serum

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