Endoscopic balloon dilatation of primary obstructive megaureter: method standardization and predictive prognostic factors

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Abstract

The management of congenital primary obstructive megaureter (POM) is usually conservative, especially during the first year of life. Endoscopic high-pressure balloon dilatation (EHPBD) is indicated when symptoms, increasing dilatation and progressive renal damage are recorded, particularly in children younger than one year of age. We identified and described endoscopic prognostic factors predicting the success or failure of endoscopic dilatation.

Thirty-eight patients (33 M;5 F) with POM from 2005-2018 were included. Diagnosis was based on US distal ureter dilatation (>7 mm), obstructive MAG-3 diuretic renogram and absence of vesicoureteral reflux (cystography). 24 patients were under 1 year of age. All patients underwent cystoscopy and high-pressure balloon dilatation with 3,5 Fr dilating balloon, after ascending pyelography. Median follow-up was of 6.5 years.

We identified characteristics with poor prognosis: stenotic punctiform ureteral ostium and/or ostium located in a bladder diverticulum (9 pts) and stenotic tract longer than 1 cm (5 pts). The patients with a stenotic tract shorter than 1 cm (18 pts) were divided into two groups: <5 mm (5 pts) and between 5 and 10 mm (13 pts) showed a good response to dilatation.

Endoscopic evaluation of ureteral ostium with pneumatic dilatation when possible is a useful diagnostic and therapeutic solution for POM treatment, especially under one year of age. EHPBD is effective in short stenotic tracts (<5 mm). It may also be repeated with good results in intermediate stenotic sections (5 mm-1 cm). According to our preliminary results, the procedure is more effective if performed earlier (3-7 months of life). Greater cohort and longer follow-up are needed to verify the stability of these results.

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Introduction

A retrovesical ureteric diameter greater than 7 mm from 30 weeks of gestational age onwards is abnormal and should be investigated postnatally.[1] In 1923, Caulk[2] introduced the term megaureter or megaloureter. Smith[3] described four types of megaureters: non-obstructed/non-refluxing; obstructed; refluxing; obstructed and refluxing. King[4] later subdivided megaureters into primary and secondary. Primary obstructive megaureter (POM) is due to abnormal peristalsis of the distal ureteral part that creates a functional obstruction (adynamic ureteral segment). The cause of this condition is due to a delay in smooth muscle differentiation. This process may last 2 years, explaining the possible spontaneous resolution of POM in approximately 80% of cases.[5] For this reason, most of megaureters are treated conservatively, especially in children under one year of age. Surgical management is required when symptoms, increasing dilatation (with progressive hydroureteronephrosis), urinary tract infections (UTI) and progressive renal damage with loss of function are recorded, in particular when initial differential renal function (DRF) is less than 40% and when conservative management fails. However, the surgical treatment of POM in the first year of life remains controversial. Before 1998, the only available surgical procedure was ureteral reimplantation, with resection of the distal ureteral stenotic end, with or without remodeling (tapering), depending on the distal ureteral diameter.[6] In newborns and children younger than 1 year this procedure could be very technically demanding and potentially threatening for bladder functional development, with high complication rate. Temporary urinary diversion may be beneficial in these cases: internal diversion with double-J stent endoscopic insertion is preferable; when not possible, external diversion (cutaneous ureterostomy) is the alternative, but it requires two or more procedures for correction.[7,8] With the advent of minimally invasive surgery, in 1998, Angulo *et al*. published the first report of endoscopic balloon dilatation for POM in children and, since then, several publications showed that the procedure is feasible, safe and less invasive in very young patients.[7-10] The endoscopic approach avoids traumatization of the bladder and preserves the blood supply of distal ureter,[11] especially in the first years of life. The recent literature focused on long-term effectiveness of EHPBD, suggesting that this procedure could be a valid option as a definitive treatment of POM. Analyzing our case series, we conclude that a correct anatomical evaluation of vesi-

coureteric junction (VUJ), ureteral ostium size and site, and length of stenotic tract represents an important prognostic factor predicting the success or failure of endoscopic dilatation and allows dividing patients into prognostic groups. In our experience, this evaluation is possible with cystoscopy and intraoperative retrograde pyelography.

Materials and Methods

Casenotes were retrospectively reviewed and data collected for patients with diagnosis of POM between 2005 and 2018 in our Center (San Bortolo Hospital, Vicenza). Diagnosis of POM was based on the following parameters: dilatation of the distal ureter greater than 7 mm at US, worsening hydroureteronephrosis at US, obstructive pattern on MAG-3 diuretic renogram and absence of vesicoureteral reflux (VUR) at cystography. Ultrasound was used to evaluate the diameter of renal pelvis and distal ureter, and the characteristics of renal parenchyma. The degree of hydroureteronephrosis was defined according to the guidelines of the Society for Fetal Urology. A voiding cystourethrogram (VCUG) was performed in all patients to rule out VUR. Antibiotic prophylaxis was not administered when diagnosis of POM was made and VUR excluded. Diuretic renogram was performed according to the guidelines of the Society of Nuclear Medicine. Good renal drainage 30 minutes after 99mTc-mercaptoacetyltriglycine (MAG-3) injection was regarded as absence of obstruction. If delay in excretion was detected, furosemide (1 mg/kg) was administered intravenously, and total urinary drainage was calculated in the following 20 minutes. A diuretic T ½ >20 minutes after furosemide injection with the persistence of more than 50% (especially in ureteral area) of the tracer was classified as obstruction. After diagnosis of POM, conservative management was undertaken, with urine cultures in case of suspected UTI, ultrasound every three months, and eventually MAG-3 renogram at 12 months.

The indications for surgery were due to clinical conditions and instrumental findings, with at least one of the following conditions: i) initial differential renal function (DRF) <40% with an obstructive excretion pattern at MAG-3 renogram or 10% of DFR loss of function related to follow-up; ii) progressive increase of hydroureteronephrosis at US; iii) febrile urinary tract infections (UTIs) not controlled by antibiotic prophylaxis.

Patients included in the protocol underwent cystoscopy under general anesthesia, and a single dose of antibiotic prophylaxis (usually amoxicillin-clavulanate 25 mg/kg) was administered before the procedure. We used an 8-9.5 Fr cystoscope with a 4-5 working channel. The first evaluation was about ureteral ostium in term of size, site, morphology and the possibility of cannulating it with a 3-4 Fr flexible guidewire. After passing vesicoureteric junction with the guidewire, a 3 Fr ureteral catheter was introduced and an ascending pyelography was performed before dilatation, to confirm the diagnosis and to measure the stenosis and the dilatation. Always under fluoroscopy control and endoscopic vision, the cystoscope was positioned in contact with ureteral ostium, allowing identifying it on radiological monitor. The length of the stenotic tract was directly measured on radiological monitor which elaborates images with a 1:1 size, starting from cystoscope edge to the end of the stenotic tract highlighted by the previously injected contrast medium (Figure 1).

We introduced on the guidewire a semi-compliant PTA dilatation balloon catheter with a size ranging from 3.4 to 5.8 Fr, a nominal diameter ranging from 2 to 10 mm and a length of 2 cm (Coyote™ES). When the balloon was located across the VUJ, it was filled with radiologic contrast medium till its nominal pressure (from 12 to 14 atm) was reached. This procedure is performed under endoscopic and fluoroscopic control and the balloon was left in place for 3-5 minutes, until the release of the stenosis became evident on fluoroscopy as the disappearance of the engraving on the balloon. Under one year of age only balloons with a maximum diameter of 8 mm were used. When dilation was completed, the cystoscope or a ureteral catheter was introduced through the distal ureter to assess the VUJ. After the procedure, we didn't leave in place nor a bladder catheter neither a double-J stent. An exception was made in two patients with solitary kidney, who had a stent positioned after EHPBD. Both patients had also a nephrostomy, which was positioned in the first days of life to reduce the pressure inside the renal pelvis. In these cases, antibiotic prophylaxis was maintained both for the stent and for the nephrostomy. The follow-up of this specific subset of patients was more strictly and included renal ultrasound at 1, 3, 9 and 12 months and VCUG in case of febrile UTI. The success of the intervention was defined by improvement of hydroureteronephrosis.

When cystoscopy showed a stenotic ureteral ostium not passable by the guidewire, or when the ostium was inside a bladder diverticulum, we suspended the procedure without performing any endoscopic dilatation. In this case, patients were observed until 1 year of age and, if clinical conditions and MAG-3 renogram are stable, they underwent open vesicoureteric reimplantation. In case of clinical or functional deterioration, patients underwent a temporary external urinary diversion before reimplantation.

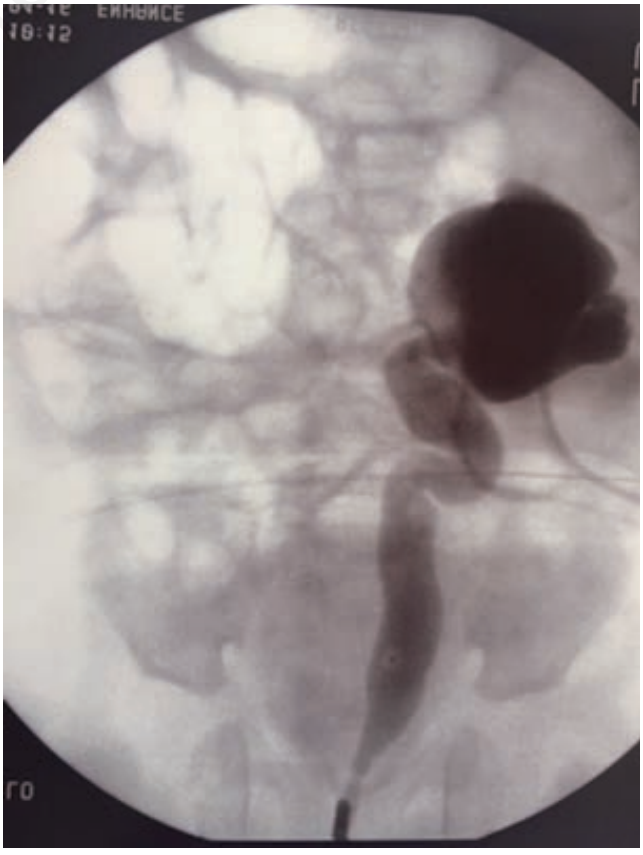

Figure 1. Endoscopic balloon dilatation.

Results

A total of 38 patients were included in the present study, 33 boys (86.8%) and 5 girls (11%), with a post-operative follow-up of at least 1 year. We excluded from the study 3 patients with a follow-up of less than 6 months. All patients had prenatal diagnosis of hydroureteronephrosis and presented with an obstructive pattern at postnatal diagnostic procedure. The most affected side was the left one, in 24 pts (63%), while right side was involved in 15 pts (35%); no bilateral cases were recorded, and two patients had solitary kidney with POM. Median age at surgery was 23 months (range: 3 months - 5.5 years) and 24 pts were younger than 1 year old. Median operating time was 20 minutes (range: 10-60 minutes). Median hospital stay was 1 day (range: 1-2 days). All patients had hospital admission of 24h, except for the two patients with solitary kidney that were discharged 3-4 days after EHPBD. Median follow-up was 6.5 years (12 months - 14 years) and no patients were lost at follow-up.

EHPBD was performed in 23 patients out of 38. In 9 cases, EHPBD could not be performed because of the failure of the guidewire to pass through the VUJ; all these patients underwent a successful open ureteral reimplantation (Cohen); in two of them, a paraureteral diverticulum was found. In other 6 patients EHPBD was not performed for the intraoperative finding of a not stenotic ostium and the absence of stenosis at retrograde pyelography. Four of them (10%) showed a spontaneous resolution (at ultrasound follow-up) of their condition, while two (5%) required a ureteral reimplantation for an associated VUR.

EHPBD was thus performed in 23 patients. All these patients presented with a stenotic distal ureteral tract; they were divided into three groups on the base of the stenotic tract length: 5 pts with stenosis <5 mm long (Group 1); 13 pts with stenosis from 5 to 10 mm long (Group 2); 5 pts with a stenosis >10 mm long (Group 3).

Group 1 (<5 mm): 4 pts presented resolution after 1 EHPBD, while 1 required a second treatment;

Group 2 (5-10 mm): 4 pts presented resolution after 1 EHPBD and 7 pts after 2 EHPBD; 1 patient resolved stenosis after a third endoscopic treatment but developed VUR; 1 patient required vesicoureteric reimplantation for worsening hydronephrosis;

Group 3 (>10 mm): 3 pts, after evaluation and EHPBD, underwent Cohen reimplantation; 2 pts required initially a cutaneous ureterostomy for their small age, followed by Cohen reimplantation after the first year of life.

The two patients with solitary kidney (a male and a female) had a stenotic tract length of 5-10 mm and both reached the resolution after one and two treatments respectively. A double-J stent was left in place and removed after 1 month from EHPBD. No stent related UTIs were recorded. There were no intraoperative complications. Retrograde uretero-pyelography through a narrow meatus never produced mucosal inflammation, swelling or bleeding that may complicate the subsequent procedure. In our series, a second endoscopic dilatation was performed if obstruction persisted, while vesicoureteric reimplantation was performed if the second dilatation was unsuccessful, relating to the length of the stenotic tract. In one case only, a third cystoscopy was performed and a calibration of VUJ done, inflating the balloon catheter at lower pression. This patient developed a postoperative 2° VUR, which was treated with Deflux™ (dextranomer copolymer in hialuronic acid) subureteral endoscopic injection. After EHPBD, follow-up included ultrasound at 3, 6 and 12 months and MAG-3 renogram at 18 months. The success was defined by the improvement in the degree of hydroureteronephrosis, the absence of UTI and the stabilization of renal function.

We didn't find any differences in terms of success of the procedure between patient younger than 1 year and older ones. In no responder youngest patients (<1 year old), the further therapeutic approach was external urinary diversion.

Discussion

Cystoscopy and ascending pyelography allow an accurate anatomical evaluation of the vesicoureteric junction in association with the measurement of the stenotic tract; this evaluation identifies anatomical and functional characteristics that are useful to establish prognostic criteria for prognosis of MOP, in terms of spontaneous resolution, successful endoscopic treatment or the need for surgical choice.

On the basis of our case study, we found the following situations.

Endoscopic dilatation is not suitable for patients with tight ureteral ostium stenosis that does not permit cannulation or ureteral ostium located within a bladder. These patients require primary surgical management, with temporary urinary diversion and subsequent vesicoureteric reimplantation as soon as possible. Likewise, patients with a stenotic tract longer than 1 cm, who don't respond satisfactorily to endoscopic treatment, need surgical treatment that depends on the age of the patient.

Patients with short stenotic tracts (<5 mm long) are successfully treated by endoscopic dilation without the need for further surgery. Patients with a stenotic tract between 5 and 10 mm belong to an intermediate prognosis group in which improvement can be achieved with repeated endoscopic treatments. In this subset of patients, at some instances the results could be definitive, while in other cases dilation allows time gaining for performing vesicoureteric reimplantation at an adequate age, avoiding temporary ureteral diversion.

Conclusions

The great part of megaureters are treated conservatively, especially in children under one year of age, because they resolve spontaneously in approximately 80% of cases, without affecting renal function. Nonetheless, 20% of cases show a worsening hydroureteronephrosis, with infectious complications and/or deterioration in renal function. In these patients an operative treatment is mandatory, even in the first months of life. Notwithstanding, the treatment of patients with POM in the first year of life is currently a challenge, both in the prevention of renal damage and in the choice of the least invasive technique available. It is also difficult to assess predictive factors for renal function loss, a parameter that universally guides therapeutic choice.

The development and affirmation of minimally invasive surgery in pediatric age in the last years led to less aggressive procedures for the surgical treatment of POM, such as the laparoscopic, robotic, or endourological approach.[7,8,11-13] Several authors described the placement of double-J ureteral stent as a temporary internal diversion in the management of POM, with good outcomes and without the need for other procedures, but with an associated high rate of comorbiditie.[7-14] Garcia-Aparicio reported a 5.9% of UTIs (4 patients) for all types of urological surgery requiring a double-J stent positioning after the procedure (laparoscopic pyeloplasty, high-pressure balloon dilatations of VUJ for POM, distal ureteral obstruction after endoscopic treatment for VUR).[15]

Christman[11] used double-J stenting after endoscopic balloon dilation with or without laser incision. Bujons[16] and Garcia-Aparicio[17] reported an absence of UTI cases after HPBD. In 1998, Angulo *et al.* published the first report of endoscopic balloon dilatation for POM in children and, since then, several publications have shown that the procedure is feasible, safe and less invasive in very young patients.[9]

On our preliminary experience, it seems possible to evaluate structural features that identify patients with poor prognosis. These features include an ostium placed in a bladder diverticulum or with a very tight diameter, and a stenotic tract longer than 1 cm: in our case series, this subgroup of patients has in fact requested a surgical approach. For short stenotic tracts, the endoscopic dilation proves to be more effective, with the possibility of repeating the procedure in the intermediate stenotic tract (5 mm - 1 cm) group. This group is the most variable in terms of response to treatment, underlining therefore the need for a personalized assessment for each case.

In our opinion, the data of the present study are very useful to choose the most suitable treatment strategy according to the anatomical and functional features of each single patient. The evaluation of the length and the aspect of the stenotic tract is easy during endoscopy and allows a better guidance to balloon dilation procedure.

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