



Primary Obstructive Megaureter: Endourological Treatment

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J. M. Angulo, A. Parente, B. Fernandez-Bautista,
L. Burgos, and R. Ortiz

62.1 Introduction

Most patients with primary obstructive megaureter (POM) only need conservative management since functional obstruction resolves spontaneously during the first months of life without renal function impairment or appearance of symptoms [1]. Surgical treatment is then reserved for those cases that develop progressive hydro-ureteronephrosis with urinary tract infections (UTI) and/or renal loss of function. However, its management and surgical options remain controversial. Ureteral reimplantation with or without ureteral tapering has been considered the gold-standard procedure for these patients, but in small infants, reimplantation of a huge ureter is challenging and leads to potential complications [2].

Endoscopic balloon dilation (EBD) of the vesicoureteral junction (VUJ) was first described by Angulo et al. in 1998 as initial approach of complicated POM [3]. Since then several publications have shown that EBD is feasible, safe, and a less-invasive procedure in the initial management of POM even for very young patients [4–7].

In recent years the interest has been focused on the long-term effectiveness of this procedure, being reported good outcomes that maintain in time, suggesting EBD as a valid option for definitive treatment in POM [8–10].

In 2004 we established in our institution the EBD of the VUJ and temporary stenting as first surgical treatment in POM with surgical criteria. In this chapter we describe our experience with this technique, its results, its complications, and its outcomes after 100 treated cases.

62.2 Patients and Methods

One hundred of POM in 92 consecutive patients were treated by EBD between years 2004 and 2016. A total of 79 POM in 73 patients (6 patients had bilateral POM) with more than 18 months of follow-up after treatment were retrospectively analyzed.

Diagnosis and management of POM were done according to the European guidelines and consensus statement of this entity. Primary obstructive megaureter was considered in those that presented progressive hydro-ureteronephrosis with distal ureter diameter greater than 10 mm, obstructive pattern on MAG3 renogram scan, and absence of vesicoureteral reflux on cystography. Nevertheless, not all of these patients needed surgical repair (in our series only 13% of cases prenatally diagnosed). The indication for surgical

J. M. Angulo (✉) · A. Parente
B. Fernandez-Bautista · L. Burgos · R. Ortiz
Pediatric Urology Unit, Department of Pediatric
Surgery, Hospital General Universitario Gregorio
Marañón, Madrid, Spain
e-mail: josemaria.angulo@salud.madrid.org

intervention was established in those with one or more of the following conditions (Table 62.1):

- Breaking through febrile UTI in 30 cases (38%) despite antibiotic prophylaxis, with clinical scenario of pyonephrosis and sepsis in 6 patients at time of treatment
- Progressive worsening of hydro-ureteronephrosis with renal parenchyma thinning in 29 cases (36.7%)
- Impairment of renal function (differential renal function less than 40% at diagnosis or decreasing more than 10% during expectative surveillance) in 20 cases (25.3%)

Table 62.1 Indications for surgical treatment

	Number of cases
UHN worsening + UTI	30 (38%)
UHN worsening with renal parenchyma thinning	29 (36.7%)
UHN worsening + impairment of DRF	14 (17.7%)
UHN worsening + UTI + impairment of DRF	6 (7.6%)
	79 POM

62.2.1 Technique

Under general anesthesia and with antibiotic prophylaxis, a cystoscopy with a 9.5 FG *Storz* cystoscope with 5F working channel is done. For some early cases of the series, we then performed retrograde pyelography before the dilation, using contrast through a 3 FG ureteral catheter.

A hydrophilic guidewire (0.014" *Choice PT™*, *J-tip*, *Boston Scientific*) or (0.018" *Radiofocus® Terumo*) is introduced through the VUJ, followed by the dilating balloon. The balloons used were semi-compliant dilation catheters with a size of 3.1 F and a nominal diameter from 5 to 7 mm and 2 cm length (*RX Muso™*, *Terumo*). Then, the balloons are filled with radiologic contrast with their nominal pressure (14 atm) with a pressure inflation device, under direct and fluoroscopic control until the complete release of the stenosis. Figure 62.1 illustrates the typical endoscopic and radiology sequence of dilation images.

When successful dilation is done, the cystoscope is introduced through the distal ureter to assess the UVJ, and a double-J stent is left in situ

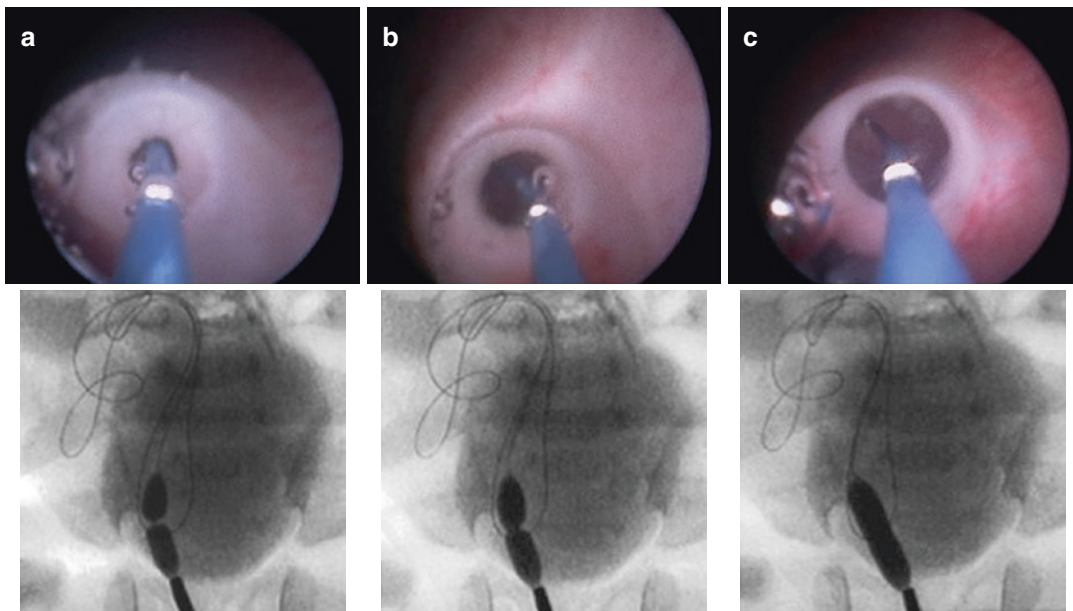


Fig. 62.1 Balloon inserted through right VUJ, endoscopic view and radiographic control. (a) Initial balloon inflation with the presence of stenotic ring; (b) progressive

dilation; (c) complete expansion of the balloon and disappearance of the stenosis

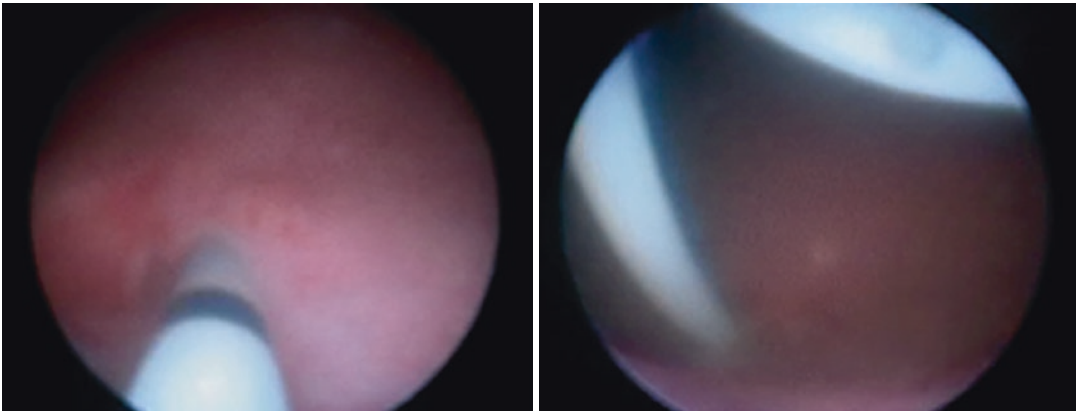


Fig. 62.2 Double-J stent placement after EBD of the VUJ

(3 Fr, 8–12 cm long, *Sof-Flex Multi-Length Ureteral Stent*, Cook Medical Europe™). After the procedure, a bladder catheter is placed during 24 h to prevent complications (Fig. 62.2).

Double-J stents are removed at 4–6 weeks at a second cystoscopy. At this time the VUJ is calibrated by distal ureteroscopy. When the cystoscope could be introduced through the VUJ, it is considered a satisfactory result. If not, a new balloon catheter is introduced and inflated to its nominal diameter to assess the VUJ diameter.

After several years performing this technique, we have done some modifications in order to achieve an easier and shorter procedure, avoiding unnecessary radiation in the majority of cases. Performing the retrograde ureteropyelography may be challenging due to the narrow ureteral meatus and may result in mucosal inflammation, edema, or bleeding. For these reasons in the last years, we are performing the balloon dilatation without fluoroscopic control, only under cystoscopic vision. We then reserve retrograde pyelography and fluoroscopic guidance for those cases in which we want to check the upper urinary tract anatomy, when dilation is being difficult or when we have problems placing the double-J stent. In the same way, we actually don't try to reach the renal pelvis with the guidewire and the double-J catheter, which is left in the dilated ureter. Overcoming ureteral loops may be technically demanding and time-consuming and needs unnecessary radiation exposure for the baby.

62.2.2 Follow-Up

All children underwent a standard follow-up protocol after endoscopic treatment; this included a clinical review and US at 3, 6, 12, and 18 months and then annually and a MAG-3–furosemide renogram scan at 6 and 18 months. Voiding cystourethrography (VCUG) was performed only if patients presented UTI or persistent ureterohydronephrosis without obstruction at the renogram (Fig. 62.3).

62.3 Results

Median age at surgery was 4 months (0.5–44), with median operating time of 20 min (10–60) and median hospital stay of 1 day [1–7]. All patients had hospital admission of 24 h except three patients in whom the endoscopic approach was done at time of urinary sepsis with ureteropyonephrosis, requiring further medical assistance after the procedure.

There were no intraoperative complications in 75 cases (94.9%). In the remaining 4 patients (5.1%), EBD could not be performed because of failure of the guidewire to pass through the VUJ in two cases (requiring open ureteral reimplantation) and unsuccessful dilation with false path in the other two cases (requiring temporary nephrostomy and posterior ureteral reimplantation).

Early perioperative complications occurred in 6 cases (7.8%). Febrile UTI after endoscopic

procedure or after double-J stent removal was reported in 5 (Clavien-Dindo 1). One patient presented ureteral double-J stent migration and developed early severe restenosis with pyonephrosis, requiring initial nephrostomy (Clavien-Dindo 3) and ureteral reimplantation weeks later.

Looking at US findings in patients who had successful initial endoscopic treatment (74/79),

significant differences were observed in distal ureteral diameter before treatment, 15 mm range (10–23); at first postoperative US after endoscopic dilation, 10 mm (0–21); and in long term, 5 mm (0–22) ($p < 0.001$ Wilcoxon test).

All patients had significant improvement in hydro-ureteronephrosis ($p < 0.05$ T-test) except those who developed restenosis or high-grade secondary VUR during long-term follow-up. Initial renal function was preserved in all patients, with normalization of the renogram elimination curves.

Postoperative secondary VUR was found during long-term surveillance in 17 cases (23%), being diagnosed in 12 after UTI and 5 after VCUG control for contralateral reflux. Subureteral endoscopic injection of *Deflux*TM (dextranomer copolymer in hyaluronic acid) was successful in 13 patients (76.4%) and failed in 4 (23.6%) who finally needed ureteral reimplantation.

Long-term restenosis occurred in 9 cases (12.2%). A new EBD procedure was successfully done in 8 cases (88.9%) at a median postoperative period of 9.5 months (5–63). Only one patient developed recurrent restenosis and finally required ureteral reimplantation.

Endoscopic approach of POM including endoscopic balloon dilation of the VUJ and endoscopic management of 2° VUR had a long-term success rate of 87.3% (69/79) with a median follow-up of 5.6 years (1.5–13.5). Endoscopic management of POM failed in 10 cases (12.7%) that finally required ureteral reimplantation (see Figs. 62.4 and 62.5).

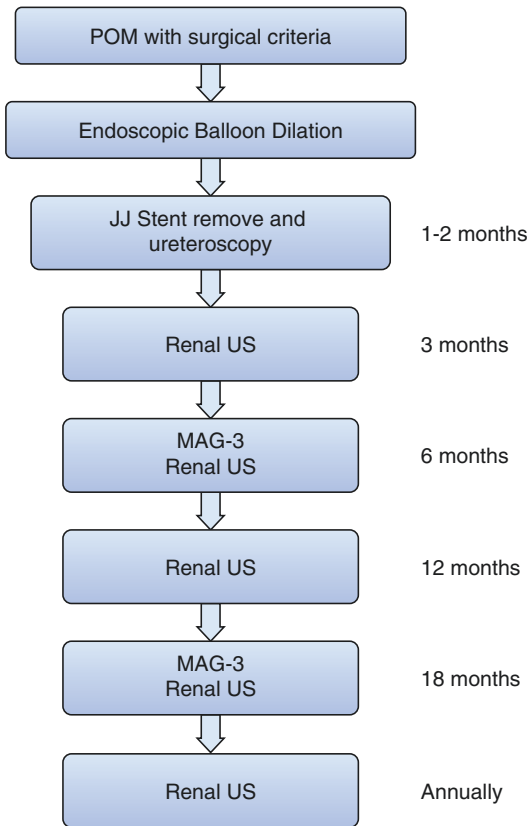


Fig. 62.3 Follow-up protocol

Fig. 62.4 Successful endoscopic management of POM

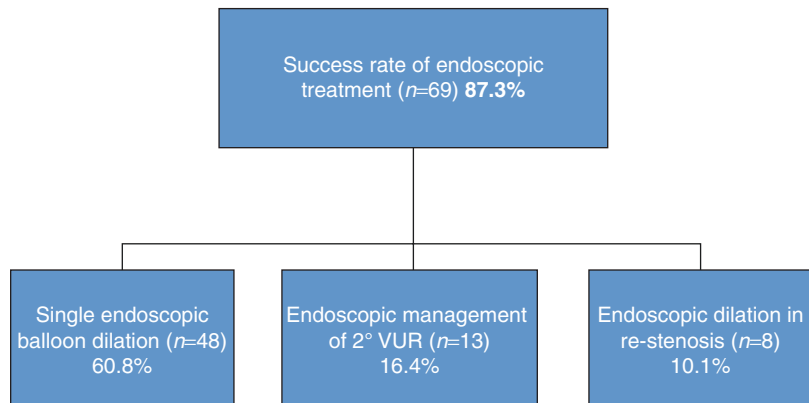
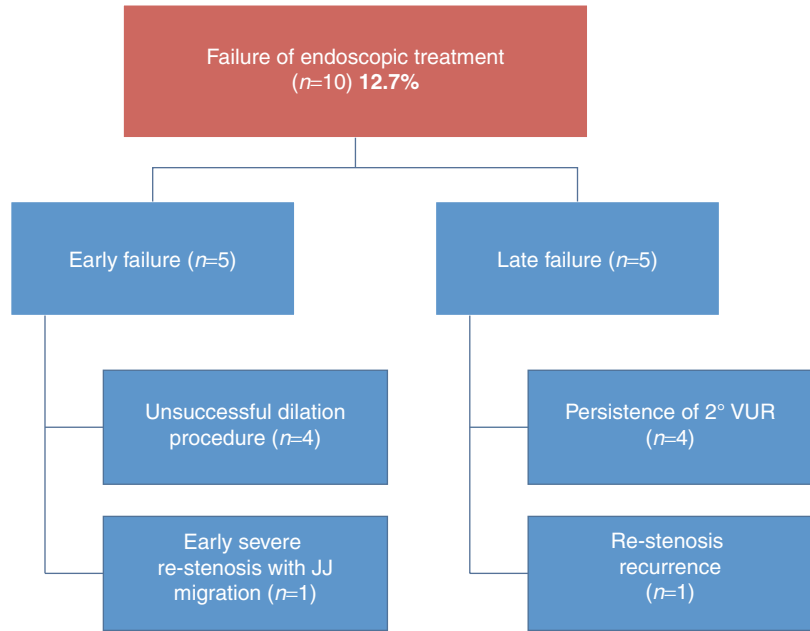


Fig. 62.5 Endoscopic failure in the management of POM



If we obviate secondary VUR and focus on the final result of EBD as treatment for ureteral obstruction, the long-term result for normalization of ureteral drainage and preserving renal function was 92.4% (73/79).

In 12 cases an ipsilateral paraureteral diverticulum coexisted with the POM. Ten of them were successfully treated by EBD showing good outcomes in long term; nevertheless, ureteral reimplantation was required in two cases (one persistent VUR and the case of recurrent restenosis).

62.4 Discussion

It is well known that POM resolves spontaneously in more than 70% of cases without impairment in renal function. However, there is a small group of patients who are going to present a progressive hydro-ureteronephrosis worsening with appearance of infectious complications and/or deterioration in renal function. These patients benefit from surgical treatment, which is usually indicated in the first months of life [1, 11].

Ureteral reimplantation with or without ureteral tapering is considered the gold-standard

procedure for these patients, with a well-documented success rate between 90 and 95%. However, reimplantation of a grossly dilated ureter in a small infantile bladder could be challenging and leads to potential complications such as secondary obstruction, vesicoureteral reflux, and bladder dysfunction. For this reason temporary urinary diversions could be indicated during first months of life, but are not exempt of complications. External ureterostomies may present problems such as infections, skin irritations, and stenosis [12]. In addition parental tolerance is usually low, demanding early closure. Percutaneous nephrostomies could be done with external tubes but have limited durability in small infants. Internal urinary diversions have become popular as proposed by Lee and Kaefer [13] who perform a refluxing megaureter reimplantation through a small laparotomy during the first months of life. However, it remains a non-definitive open surgery and creates a high-grade secondary VUR.

The important development of minimally invasive techniques achieved in pediatric age in the last years has led to nonaggressive procedures for the surgical treatment of POM such as the laparoscopic, robotic, or endourological approach. Nevertheless, we cannot obviate that

the main objective of any technique even minimally invasive must be to obtain similar outcomes to the gold standard or at least good results with less morbidity or complications.

Several authors have postulated the placement of double-J ureteral stent as a temporary internal derivation in the initial management of POM, with good outcomes in a group of patients that did not need any more procedure but controversial results and remarkable comorbidity in an important number of cases.

Since endoscopic balloon dilation was first described by Angulo et al. [3] as an initial treatment for children with complicated POM, several publications with few patients and short follow-up periods showed that EBD using the original technique or variations of the same principle was a feasible, safe, and less-invasive procedure for the initial management of POM with surgical criteria even for very young patients. In 2007 Angerri et al. [4] reported their initial experience with six patients in whom urinary obstruction disappeared without associated complications in a median follow-up of 31 months. Christman et al. [5] reported in 2012 their experience after the treatment of 17 children with a follow-up of 3.2 years. These authors added a laser incision in cases of ureteral stenosis greater than 2 cm and placed two double-J stents in the ureter simultaneously, reporting good long-term outcome with disappearance of hydro-ureteronephrosis in 71% of the series. García-Aparicio et al. [6] presented a series of 13 patients with a medium-term success rate of 84.6% (11 of 13), requiring ureteral reimplantation in 3 patients (2 persistence of UHN and 1 high-grade VUR).

Recent publications have focused on establishing long-term effectiveness of EBD as definitive treatment of POM, confirming good results with minimal associated morbidity. Romero et al. [8] reported in 2014 the experience of our institution in 29 patients treated until 2010, with a median age at treatment of 4 months and a median follow-up of 47 months. It was concluded that the patients who had a favorable evolution with disappearance of the UHN and adequate renal drainage confirmed by renogram remained asymptomatic and with stable situation during

the subsequent follow-up. Five patients had secondary VUR and three of them were satisfactorily treated endoscopically. Finally, the endourological management of the POM including EBD of the VUJ and treatment of 2° VUR had a success rate of 86%. Bujons et al. [9] have reported excellent results in 19 patients, with a long-term success of 90% after the initial dilatation procedure and a follow-up of 6.9 years. One patient required a second dilatation due to restenosis and another one endoscopic treatment of 2° VUR, both with good outcome. Casal et al. [10] have just communicated good outcomes in a short series of 13 patients but with an important median follow-up of 10.3 years (4.7-12-2), asserting the value of balloon dilation as a definitive treatment for POM.

Technical variations to the initial procedure have been proposed with encouraging results. The group of Kajbafzadeh [14] reported in 2007 a long series of patients treated by endoureterotomy (ureterotomy and detrusorotomy at 6 h) leaving double-J stent for 1 week, without associated comorbidity and with a complete resolution of ureterohydronephrosis in 71% of cases. Capozza et al. [7] published the dilation of the VUJ with Cutting Balloon™ in three patients with persistence of the stenotic ring during the previous endoscopic high-pressure balloon dilation, obtaining a complete resolution of the stenosis and good postoperative course.

Even the advantages described of EBD, the endourological management of POM remains controversial. The aspects to be discussed focus on secondary VUR, the possibility of restenosis, and the use of radiation in young patients. Additionally, it is difficult to assess its value as a definitive treatment in POM attending to the short experience reported in the literature.

Regarding secondary VUR, García-Aparicio [15] analyzed it in his group of patients, reporting 27% (6 cases of 22 POM treated). Of these, two were treated endoscopically, and two were treated by ureteral reimplantation. The author concluded that the coexistence of ipsilateral paraureteral diverticulum is a risk factor for developing secondary VUR; however the number of cases was very low (two of four) to establish a reasonable

conclusion. In the series published by Bujons et al. [10], only 1 case of 19 presented secondary VUR, and it was resolved endoscopically.

In our series secondary VUR was found during long-term surveillance in 17 cases (23%). Endoscopic treatment of it was successful in 13 patients (76.4%) and failed in 4 (23.6%) who required ureteral reimplantation. For these patients with 2° VUR, three had an ipsilateral para-meatal diverticulum and only one required reimplantation. In our experience, the presence of para-meatal diverticulum was not a bad prognosis factor for the endoscopic management of POM, since 10 of 12 cases of the series had an excellent outcome.

Long-term restenosis occurred in 9 cases of our series (12.2%). A new EBD was done with good long-term outcome in 8 cases (88.9%) till the date. Only one patient developed recurrent restenosis and finally required ureteral reimplantation. The role of Cutting Balloon™ dilation may be a useful option in these cases. We used it recently with excellent midterm outcome in three patients treated at other institutions who developed restenosis after initial EBD of the VUJ. Then, we actually reserve the Cutting Balloon™ dilation for future restenosis or in primary cases when the stenosis is not completely solved with the balloon catheter at time of initial EBD.

Attending to our experience and looking at the literature, we can consider EBD of the VUJ as a relatively simple technique, reproducible, and with a short learning curve compared to other procedures. However, its success lies in the use of adequate endoscopic material. Appropriate hydrophilic guidewires (0.014"–0.018"), balloon catheters with low profile (2.7CH), and double-J stents suitable for pediatric age are crucial both for the success of the technique and to avoid complications.

62.5 Conclusion

Endoscopic balloon dilation has shown to be a safe, feasible, and really less-invasive procedure in primary obstructive megaureter with surgical criteria even in small infants.

In our experience we can consider it an effective treatment with few postoperative complications and good outcomes that maintains at long-term follow-up. The main complication observed was secondary VUR; notwithstanding it did not result in significant morbidity for the patients and could also be treated endoscopically with a high success rate.

In comparison with the conventional surgery, EBD has the obvious advantages of being a minimally invasive procedure, with a shorter operating time, immediate recovery, and with no patient-age limitations. In our opinion, it may be considered first-line treatment in the management of POM in children, avoiding unnecessary bladder surgery in the vast majority of patients. Nevertheless, it doesn't invalidate ureteral reimplantation in case of failure.

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