

Guidelines of the Italian Society of Videosurgery in Infancy for the minimally invasive treatment of the ureteropelvic-junction obstruction

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Background

The SIVI (Italian Society of Videosurgery in Infancy) guidelines are clinical practice guidelines edited and approved by the Society's steering committee. They are the products of a detailed systematic review of the literature, integrated with expert opinion in the field of pediatric minimally invasive surgery.

These guidelines are intended to assist the pediatric surgeons experienced or not experienced in minimally invasive surgery, with the goal to inform choices related to the indications, approach, and techniques to use when treating the major pediatric pathologies.

Given the complexities of congenital malformations and other pediatric surgical conditions, as well as large variations in available regional health services, we must note that these guidelines are not intended as a cookbook recipe to follow for all possible patients. Rather, the guidelines should serve as a flexible frame-

work, to be used by the physician in concert with the parents, to choose the best approach for each individual patient. Decisions tailored to available scientific knowledge and the needs and desires of the patient's family serve both patient autonomy and medical science.

All guidelines are published in this scientific Journal, in order to ensure their availability to all physicians.

The Guidelines project has been approved by the SIVI General Assembly of the 2016 Madrid congress.

Review of guidelines has been performed by the Steering Committee of SIVI and experts.

Clinical classification and epidemiology

The hydronephrosis, characterized by the dilation of the renal pelvicalyceal system with possible functional damage to the renal parenchyma, is the most common congenital abnormality of the urinary system detected in utero through the prenatal ultrasound screening. However, nowadays it is widely known that the pelvicalyceal dilation is not necessarily a sign of parenchymal obstruction or distress. It is furthermore important to underline that approx. 60% of pyelectases that are recognized in utero tends to spontaneously resolve within the first months or years after birth. It was calculated that approximately less than 5% of cases of hydronephrosis observed during pregnancy result in a real obstruction of the ureteropelvic junction that requires a pyeloplasty. A side from the diagnosis via prenatal or neonatal ultrasound screening, the presentation symptoms in older infants are usually abdominal pain, such as lumbar pain or colic, or urinary tract infections.

Definition

The obstacle to the anterograde passage of the urine through the Ureteropelvic Junction (UPJ) normally causes an increase, sometimes progressive, of the intrarenal pelvicalyceal system, with a

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wide spectrum of variations noted for the reduction of the parenchymal function at the renal scintigraphy. It is not rare that the kidney function appears normal for a long period of time, sometimes the obstruction is so tightened that it determines the complete loss of function for the affected kidney. The etiology and pathogenesis at the base of the UPJ obstruction might vary: adynamic hypoplastic segment of the upper part of the junction and the proximal part of the ureter; valve leaflets; high insertion of the ureter in the renal pelvis; tortuous proximal ureter (corkscrew ureter); aberrant inferior pole vessels (extrinsic obstacle to the excretory duct). In 5-10% of cases of UPJ obstruction, a second abnormality of the urinary system is associated such as Vesicoureteral Reflux (VUR), Vesicoureteral Junction (VUJ) obstruction, homo- or contralateral non-obstructive and non-refluxing primary megaureter, contralateral multicystic dysplastic kidney.

Diagnosis

In the last 2-3 decades, an important update in the diagnosis of pediatric hydronephrosis has consolidated, clearly differentiating from the diagnosis of hydronephrosis in adult patients. Blood chemistry exam: the blood chemistry exams for the renal function, and in particular the creatinine, are recommended especially if both kidneys are affected. Kidney and urinary tract ultrasonography: it represents the first morphological study, regardless of the patient's age. The ultrasonography scanning performed within 3-4 days after birth could falsely reassure on the recovery from hydronephrosis recognized in utero due to the neonatal dehydration with postnatal weight loss. Therefore, it is recommended to perform an ultrasonography 4-5 days after birth, normally between the 1st and 2nd week after birth. The most useful ultrasonography parameters are the renal longitudinal diameter, the parenchymal thickness at the mid-portion of the kidney and, most of all, the anteroposterior diameter (APD) of the pelvis at the hilum. This latter is far and away the most significant parameter, that is also necessary for the follow-up. Both kidneys need to be evaluated, also in the posterior projection. Attention needs to be paid in evaluating the possible calyceal dilation and the possible presence of mono- or bilateral ureteral dilation. The presence of an homolateral dilated ureter might lead to different possible diagnoses, from UPJ obstruction, VUR, VUJ obstruction or primary megaureter.

Micturating cystourethrogram

This exam is useful to evaluate the potential presence of VUR or obstacle in bladder emptying, as in case of congenital valves of the posterior ureter. It needs to be performed under urinary antibacterial prophylaxis, with sterile urine. However, the exam is recommended if, at the ultrasonography scanning, dilated or echographically visible ureter, overdistended bladder and/or with thickened walls, duplicated ureter or solitary kidney, or hyperechoic kidneys are noted also after the first weeks after birth. Nowadays the cystography or the cystosonography are not considered necessary studies in case of isolated single hydronephrosis with no other ultrasonographic pathological findings noted for the urinary tract. The indication for cystoscopy is questionable in case of simple bilateral hydronephrosis, with no ureteral dilation or other urinary abnormalities recognizable at the ultrasonography.

Dynamic renal scintigraphy

The dynamic renal scintigraphy is a functional exam that uses the MAG3 radioisotope (marked with Technetium-99m) or, more

rarely, the DTPA, and it has become essential in the pediatric age to study the renal function and the excretory phase of the radio urine, through the evaluation of the renal radioisotope uptake and the pelvis drainage curves. In the initial phase of uptake, the parenchymal perfusion and the differential function of each kidney, that contributes in percentage to the global kidney function, are evaluated. The pelvic drainage phase is represented by a curve, whose inclination after the uptake represents the ureteropelvic drainage speed. In case of reduced or absent curve due to an altered ureteropelvic drainage, the furosemide stress test (0.5-1.0 mg/kg) is also performed: an insufficient drainage, i.e. a junction obstruction, is determined when the isotope half-time ($T_{1/2}$) in the renal pelvis region is >20 min. However, this exam partially depends on various factors such as the infant hydration, the renal function and the bladder filling, for which precise guidelines were issued by the European Society for Pediatric Nuclear Medicine. The renal scintigraphy is ideally recommended starting from the 3rd month after birth, when the renal development is completed, however, in selected cases, it can be performed starting from the 1st month after birth. The use of the technique in the neonatal period is less reliable.

Static renal scintigraphy

The scintigraphy performed with Dimercaptosuccinic Acid marked with Technetium-99m is used in case of coexisting VUR to verify the presence of renal scarring or in case of poorly-functioning unilateral kidney in order to decide to either opt for a possible nephrectomy or instead decide on a conservative renal treatment. A prompt diagnosis, by means of the above mentioned studies, is necessary in young infants with kidney with hyperechoic aspect at the ultrasonography, prenatal history of oligohydramnios, solitary hydronephrotic kidney, severe bilateral hydronephrosis ($APD >20$ mm), or abnormal bladder or with thickened walls, as in case of congenital posterior urethral valves. These conditions might predispose to progressive renal failure and to episodes of urosepsis also at the risk of the young patient's life.

Indication for surgery

The surgery to remove the UPJ obstruction is indicated in case the hydronephrosis with obstruction to the urine flow is confirmed. Traditionally, the parameters that predict the obstruction are a differential renal function inferior to 40% and/or a flat elimination curve or an increase of $T_{1/2} > 20$ min. At the ultrasonography, a progressive increase of the pelvis APD or an ADP > 30 mm with dilated calyces is considered significant (90% risk of surgical intervention). Clinical indications for surgery are also urinary tract infections and recurrent pain. Particular attention needs to be paid to the correct indication for surgery in hydronephrosis with intrarenal pelvis or with considerable calyceal involvement, in bilateral hydronephrosis and in solitary kidney.

Surgical approaches

Open surgery

The Anderson-Hynes dismembered pyeloplasty or pyeloplasty with UPJ resection is perhaps still considered the gold standard however new minimally invasive video-laparoscopic techniques are rapidly gaining ground. The technique has a success rate of 95-98%. During the first years of age, the retroperitoneal approach is

normally used with subcostal incision or posterior lumbar incision or, even better, abdominal wall muscles splitting with direct access to the renal fossa and the UPJ. The resection of the junction and eventually of the exceeding pelvis is followed by the longitudinal spatulation of the proximal end of the proximal ureter and by the ureteropelvic bevelled anastomosis with half-running or long-term absorbable interrupted sutures (caliber 5/0 or 6/0 or 7/0). The application of a ureteropelvic transanastomotic catheter is optional. Different types of stent can be used, depending on the surgeon's preference: the Double-J stent and the nephrostomy ureteropelvic stent are the most used. It is possible to apply a decompressive temporary nephrectomy with optional drainage in the renal fossa. Usually, but not necessarily, a vesical catheter helps to maintain the upper urinary tract decompressed during the first 2-3 days after surgery. This technique is nowadays often performed via minimally invasive video-laparoscopic approach. Other pyeloplasty techniques are rarely used in young patients: Scardino-Prince pyeloplasty, Foley Y-V plasty, Culp-DeWeerd and the Fenger plasty (similar to the Y-V with longitudinal incision of the junction that is transversally resutured as in Heineke-Mikulicz). The ureterocalicostomy might be used in particular conditions such as in scarring at the level of the UPJ and hydronephrosis in horseshoe kidney. A prerequisite for the success of the ureterocalicostomy is the excision of a good part of the parenchyma to allow a good anastomosis between the calyceal and proximal ureter urethelium.

Minimally invasive surgery

The minimally invasive surgery includes techniques that are recently obtaining a greater diffusion, substituting, for the most part, the traditional open techniques. It includes the laparoscopic and retroperitoneoscopic approaches to the UPJ, the one trocar assisted pyeloplasty, the laparoscopic vascular hitch procedure and the robot-assisted pyeloplasty.

Preoperative workup

The blood exams that are normally required are: complete blood count, creatinine, electrolytes, coagulation tests and, eventually, blood type. Especially for the transperitoneal laparoscopic procedures, it is recommended to perform a bowel preparation with anti-fermenting drugs and laxatives and/or an enema within 24 hours before surgery. The surgery consent form should include information on the diagnosis, the recommended surgical procedure, the alternatives, the possible need for conversion. It is recommended to inform on the type of stent and catheters that will be used and ultimately on the possible complications such as bleeding, urinary leakage, urinary infections and risk of persistent obstruction with possible need for a second surgery.

Operating room preparation for the transperitoneal laparoscopic approach (pyeloplasty and vascular hitch)

The patient is positioned on a lateral or semi-lateral position; the side opposed to the hydronephrotic one is positioned towards the edge of the surgical table. The surgeon stands in front of the patient, with the laparoscopic video tower positioned behind the patient. The laparoscopic instruments normally include: 2 needle holders, swab stick, graspers, surgical hook, mixters, aspirator-irrigator, monopolar and/or bipolar electrosurgical devices, and they can vary according to the operating surgeon's preference.

Operating room preparation for the retroperitoneoscopic approach

The patient is positioned on a lateral decubitus with a soft pillow or roll under the contralateral flank in order to widen the space

between the costal margin and the iliac crest allowing the positioning of the laparoscopic ports. The surgeon and the assistant stand behind the patient, the surgical nurse stands next to them or at the feet and the laparoscopic video tower is positioned on the other side, i.e. towards the abdomen. The laparoscopic instruments are similar to those used in the transperitoneal access.

Surgical technique: laparoscopic pyeloplasty

A 5- or 10-mm trocar is normally used at the umbilical level, eventually with balloon (Hasson trocar), for a 5- or 10-mm optic (possibly 30-degree) and two 3- or 5-mm ports for the instruments. These latter need to be positioned in order to obtain the correct triangulation of the laparoscopic instruments. Sometimes a third port might be necessary (for example to lift the liver lobe during the right pyeloplasty or to suspend the anastomosis). On the left side, more frequently a transmesocolic window is sufficient to reach the retroperitoneal space and then perform the pyeloplasty or vascular hitch. On the right side, it might be more often necessary to mobilize the hepatic flexure of colon to access the renal fossa in correspondence with the UPJ. The Anderson-Hynes pyeloplasty is then performed similarly to the open procedure, with junction resection, proximal ureter spatulation and a bevelled anastomosis. The intracorporeal suture normally requires a certain degree of laparoscopic expertise and it can be performed with two halves running or interrupted sutures, using long-term absorbable monofilament or braided threads, normally 5/0 or 6/0. The application of the internal ureteropelvic Double J stent is not mandatory however it is often used to secure the pelvic drainage during the immediate postoperative period. The stent can be inserted with various methods, generally with the anterograde insertion through a laparoscopic port or percutaneously on a needle cannula or sometimes also cystoscopically. At the end of the procedure, the peritoneal window is normally closed even though there are no supporting scientific evidences. A drainage is normally positioned in the renal fossa coming out from a slanting percutaneous counteropening or from the most distal laparoscopic port.

Surgical technique: retroperitoneoscopic pyeloplasty

In most cases, two 3- or 5-mm surgical ports and one port for the 5- or 10-mm optical system suffice. A 0- or 30-degree optical system is used according to the operator's preference. An 8-12 mm skin incision is made at the apex or below the 12th rib: the peritoneum is accessed via blunt dissection, with the psoas muscle providing guide and orientation. As the retroperitoneum is a virtual cavity, in order to develop a sufficient working space, moist gauzes or more rarely an air-inflated balloon are introduced through the first incision (the longest) following the technique introduced by Gaur. In older patients, finger dissection can be performed. A great attention needs to be paid in order to avoid the perforation of the parietal peritoneum and the following gas leak inside the peritoneum with consequent difficulty in creating the working space. Once sufficient retroperitoneal space is created, the kidney appears suspended and attached to the peritoneum upwards and the pelvis appears inferiorly with the ureter running on top of the psoas; two-three 3- or 5-mm working trocars are then introduced. Once the UPJ is bluntly isolated, the Anderson-Hynes dismembered pyeloplasty can be performed observing the already described principles and surgical timing. Normally the retroperitoneal video-laparoscopic procedure is more demanding due to the reduced working space and the need for a high-level expertise in MIS. Technically, the following steps are identified, even though they can vary according to the operator's experience and

preference: A) Junction anchoring on the psoas muscle. B) Opening of the dilated pelvis in a declive position and spatulation of the proximal ureter. C) suturing of the anterior border of the ureteropelvic anastomosis (interrupted or running sutures). D) Resection of the UPJ and of part of the redundant pelvis (not necessarily). E) insertion of internal Double J stent via anterograde percutaneous access. F) Completion of the anastomosis on the posterior border and completion of the pyeloplasty with running or interrupted suture. G) Possible drainage of the renal fossa.

Surgical technique: one-trocар-assisted pyeloplasty

This technique combines the advantages of the minimally-invasiveness of the video-laparoscopic approaches with the practicality and safeness of the open pyeloplasty at the cutaneous level. However, it can be used with increased safety in patients with reduced body weight (<20kg), no older than 5-6 years of age, with reduced fat pad and relatively thin abdominal walls. The main stages of the procedure consist of: A) 15-18 mm skin incision at the apex or below the 12th ipsilateral rib, as in the previously described retroperitoneoscopy. B) Introduction of a 10-mm Hasson balloon trocar and 10-mm optical instrument through a 5-mm surgical port. The 5-mm laparoscopic instruments are similar to the ones used in other video-laparoscopic procedures. C) Blunt mobilization of the posterior peritoneum from the psoas muscle to the renal fossa via optic instrument and laparoscopic swab, favored by a CO₂ pressure of approximately 9 mmHg. D) Identification of the lower pole of the kidney, the proximal ureter and the pelvis. E) Vessel-loop lifting of the UPJ to the subcutaneous level. F) Working trocar removal and continuation of the procedure via traditional Anderson-Haynes technique at the cutaneous-subcutaneous level (loops or optical magnification), exteriorizing the obstructed UPJ through the laparoscopic port. G) Double J stent insertion and suturing completion with 5/0 or 6/0 sutures, repositioning of the junction in the renal fossa and laparoscopic verification of absence of excretory duct kinking or torsion. H) Elective application of a drainage in the renal fossa. Useful Foley catheter for 24-72 hours. The technique does not require a high-level laparoscopic experience as intracorporeal sutures are not expected. It can also be used in case of aberrant vessels however it requires the section of the junction and the ureteral vessel transposition. Normally, it cannot be used for the vascular hitch. If necessary, the widening of the incision of the laparoscopic port can be easily performed and it is possible to proceed with a semi-conversion.

Surgical technique: vascular hitch

This technique can be performed in case of lower pole aberrant vessels obstructing the proximal ureter. In order to use this technique, a correct selection of the patient is essential: the vascular hitch is reserved to cases of hydronephrosis caused by pure extrinsic compression of the junction due to lower pole aberrant vessels. It is necessary to know that the extrinsic compression might coexist with an intrinsic compression of the UPJ: these cases need to be recognized in order to avoid a long-term failure of the eventually adopted vascular hitch technique. The technique is certainly attractive as it does not require the section and the anastomosis of the excretory duct and the non-systematic application of ureteropelvic stent. The classic shape of hydronephrosis caused by aberrant polar vessels normally occurs in school-age children or adolescents with pain and intermittent pelvic dilation more frequently without calyceal dilation. The presence of aberrant vessels is verified via Color Doppler ultrasonography and MRA, although their presence become certain only via surgical exploration. The laparoscopic transperitoneal access is the first-choice procedure as the vessels

run anteriorly to the excretory ducts and it is analogous to the one performed for the dismembered pyeloplasty. Therefore, once the aberrant vessels are identified, they are carefully displaced cranially on the pelvis in order to release the junction. An intraoperative test is performed with the administration of furosemide (1mg/kg) after water load (20mL/kg) to laparoscopically visualize the good ureteropelvic transit with pelvic decompression, therefore excluding the coexistence of an intrinsic obstruction. A difficulty in emptying the pelvis after the test, a thin junction with fibrotic and non-linear aspect should warn the surgeon in performing the vascular hitch instead of the classic Anderson-Hynes. It is possible to use two or three 3- or 5-mm ports and an umbilical 5- or 10-mm optical port. The access to the renal pelvis is analogous to the one used for the pyeloplasty. The lower pole vessels are identified, mobilized (eventually, on surgical tape) and the pelvis together with the proximal ureter are adequately released from possible adhesions. The water load test with furosemide has to demonstrate a good urine passage with pelvic decompression. In case of doubtful response, it is possible to perform a pyeloplasty with ureterovascular transposition (Anderson-Hynes dismembered pyeloplasty) or without transposition (ex. Fenger plasty). The anchoring of the polar vessels cranially and distally to the UPJ is normally realized with two/three interrupted sutures that secure the perivascular tissue to the pelvic wall as in Hellstrom procedure (1949) or by creating a hammock with the pelvic wall, to sustain the vascular pedicle, as suggested by Chapman (1959) with two/four 2/0, 3/0 or 4/0 absorbable or non-absorbable sutures. This latter appears the simplest and safest technique to avoid damages to the aberrant vascular pedicle. Although this technique is not recent and the latest data from the literature are reassuring, the long-term results of this video-laparoscopic technique are still to be confirmed and a careful follow-up is recommended.

Surgical technique: robot-assisted laparoscopic pyeloplasty

This technique is normally performed via transperitoneal approach, by using a 10-mm umbilical port for the optic and two 5-, 8- or 10-mm working ports. Occasionally, it is possible to use an additional working port. The technique is similar to the one used in the described transperitoneal laparoscopic pyeloplasty, however it preferably adapts to children older than 4-5 years of age, even though cases of application also during the first year of age are reported in literature. The great advantage of the robot assistance is due to the relative easiness in performing the intracorporeal suture. Moreover, further advantages are represented by a better ergonomics for the surgeon, a three-dimensional vision, hands tremors filtering and a greater articulation obtained by the laparoscopic instruments. The disadvantages are represented by instruments and ports that are not ideal for younger patients and, above all, by the high purchasing and maintaining costs of the robot, that are still nowadays prohibitive for some pediatric urological surgery centers.

Surgical technique: endopyelotomy

Several balloon catheters have been suggested and used for the dilation or endoscopic dissection of the UPJ, via anterior nephrectomy approach or via retrograde ascending approach, via ureteroscopy or under radiological guidance. This technique requires the application of ureteropelvic Double-J stents. A discrete success was demonstrated in adult patients with history of failed pyeloplasty, while no satisfactory results have been reported for the congenital hydronephrosis due to intrinsic junction condition in pediatric patients. This technique does not have to be used in case of suspected lower pole aberrant vessels.

Post-operative treatment and follow-up

In literature, there is a wide variety on the use or non-use, at the surgeon's discretion, of ureteropelvic stents, peripelvic drainage and vesical catheter with no certain evidence of a statistically valid efficacy. Generally, a transanastomotic stent is used: either an external nephrostomy catheter, to be removed after 7 days, or an internal Double-J stent, to be removed via cystoscopy with foreign body forceps after approx. 4-6 weeks. The renal fossa drainages are normally removed within 3-4 days, in absence of wound secretions. The antibiotic coverage is recommended during the first 7 days and a low-dose prophylaxis might be maintained in case of internal Double J stent that might cause possible bacterial colonization and reflux. The instrumental follow-up studies normally consist of a renal ultrasonography 3, 6 and 12 months after surgery and a MAG3 dynamic renal scintigraphy 6-12 months after surgery although the frequency of the follow-up studies might vary according to the surgeon's preference and the specific condition of the patient who underwent surgery. It might be recommended to periodically monitor the arterial blood pressure especially in patient with reduced residual renal function due to the risk of renovascular hypertension. It is often recommended to perform a complete nephrology/urology follow-up at puberty.

Conclusions

The standard pyeloplasty with open lateral access is a technique with a high rate of success (95-98%) in solving the upper junction obstruction. Therefore, any innovative approach, in particular the minimally invasive video-laparoscopic techniques, needs to confront with it. The minimally invasive techniques represent, without doubt, a great advantage for the young patient and for the length of hospital stay, especially in older children. An intermediate technique between the open surgery and the laparoscopic surgery is the retroperitoneal video-laparoscopic assisted pyeloplasty (OTAP-OPRAP) that might represent a valid and practical option before moving to the pure minimally invasive intracorporeal technique. The Anderson-Hynes dismembered laparoscopic pyeloplasty is more demanding, especially when performed with a retroperitoneal access, however it guarantees optimal results in experienced hands. The transperitoneal laparoscopic vascular hitch technique offers optimal results in case of hydronephrosis caused by aberrant vessels, provide that a rigorous case selection is performed. The robot-assisted pyeloplasty is optimal in young patients older than infants as long as the instruments and trained personnel are available.

What is certain is that the minimally invasive video-laparoscopic techniques are currently replacing the traditional open pyeloplasty in an increasingly number of Pediatric Urology and Surgery centers.

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