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Giant hydronephrosis in adults: What is the best approach? Retrospective analysis of 24 cases

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ABSTRACT

Background: The distension of the pelvis and calyces of the kidney due to the obstruction and stasis of urinary flow or Hydronephrosis, caused by a lesion in the upper or lower urinary tract. The giant hydronephrosis (GH) is a rare entity and its etiologies are varied. Most reported cases of GH occur in infants and children, and are congenital in origin.
Objectives: To formulate and validate a strategic approach for the treatment of giant hydronephrosis (GH) based upon anatomical and functional status of renal units in adults.
Patients and Methods: We present a retrospective review about 24 cases of GH managed between February 2001 and February 2010. Epidemiologic data, radiological investigations, therapeutic indications, preoperative findings and follow-up were reviewed. Therapeutic indications were based upon functional status of GH.
Results: The age of the patients ranged from 19 to 61 years. Ten patients were males and 14 were females. IVU revealed non-visualized unit of the affected side in 4 patients. The quantity of urine drained was between 1.1 litres and 3.5 litres. Seven patients were subjected to nephrectomy. Eight patients underwent reduction pyeloplasty. Nine patients were treated for urolithiasis. Follow-up was in the range of 9 to 73 months (mean = 32 months). Four patients had chronic renal failure requiring hemodialysis and one patient presented with recurrent pyelonephritis.
Conclusions: In very poorly functioning unit, nephrectomy is the procedure of choice. In salvageable unit, anatomical configuration should dictate the type of reconstructive procedure.

▸ *Implication for health policy/practice/research/medical education:*
This paper can be useful in approach to management of giant hydronephrosis which is a rare entity.

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1. Background

Hydronephrosis is the distension of the pelvis and calyces of the kidney due to the obstruction and stasis of urinary flow caused by a lesion in the upper or lower urinary tract (1). The giant hydronephrosis (GH) is a rare entity. Its etiologies are varied (2). It was defined arbitrarily by Sterling (3) in 1939 as a kidney containing a litre or more of fluid in the collecting system (4). Most reported cases of GH occur in infants and children, and are congenital in origin. This situation is a rare urological entity in adults

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(5) and often misdiagnosed clinically (6). Since the first description, only case reports have been published. We report a series of 24 cases of GH.

2. Objectives

Our purpose is to discuss treatment and formulate management strategy in these cases based upon etiology, location of obstruction, anatomical configuration and functional status of renal units.

3. Patients and Methods

We present our experience through a retrospective study. Between February 2001 and February 2010, 24 adults with GH were treated in our institution. Epidemiologic data, radiological investigations, therapeutic indications, preoperative findings and follow-up were reviewed. Functional value of the hydronephrotic kidney was evaluated according to scintigraphy data and renal parenchyma thickness (more than the half of all renal surface). Our treatment policy was based upon a strategic approach summarized in *figure 1.* Nephrectomy is often performed due to severe impairment of renal function. Follow-up was conducted clinically, measure of creatinine level and IVU and or renal scintigraphy every 6 months.

4. Results

The mean age of patients was 39.4 years (19-61). Ten patients were males and 14 were females with a sex ratio of 2:3. The common clinical presentation was flank pain and/or renal colic which were reported by all patients except one. All patients denied having fever and hematuria.

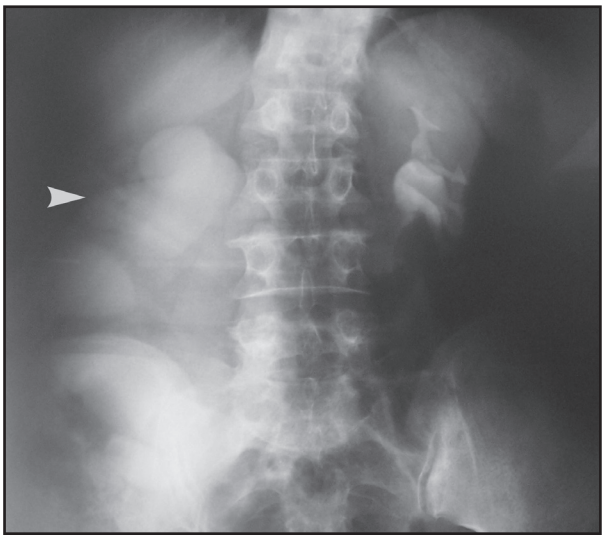

Figure 2. IVU: GH of the right kidney due to UPJ obstruction (arrow).

On clinical examination a cystic smooth lump was found occupying nearly a hemi-abdomen in 5 patients. Serum creatinine was normal in 19 patients and slightly high in 3 patients. Two patients presented with azotemia necessitating hemodialysis. Ultrasonography, performed in 19 patients, revealed bilateral massive hydronephrosis in 10 patients and normal contra-lateral unit in the others. Intravenous urography (IVU), performed in 20 patients *(Figure 2)*, revealed non-visualized unit on affected side in 4 cases. CT scan was performed in 6 patients *(Figure 3)*. Renal scintigraphy, performed in 16 cases, concluded that the functional kidney participated less than 20% of the total renal function in only 6 of them. GH was in the right side in 14 patients and bilateral in one case.

Plain X-ray KUB and IVU revealed a calculus in the renal

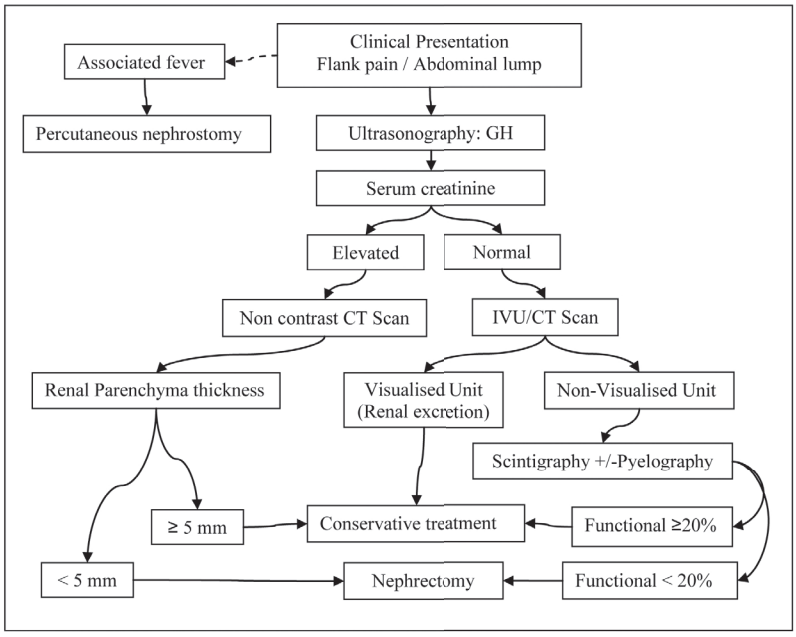

Figure 1. Strategic approach for management of GH

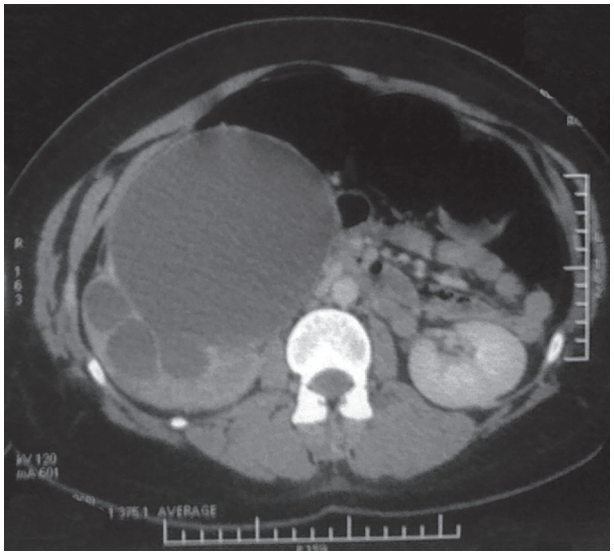

Figure 3. CT scan: A giant cystic mass occupying the right side of the abdomen

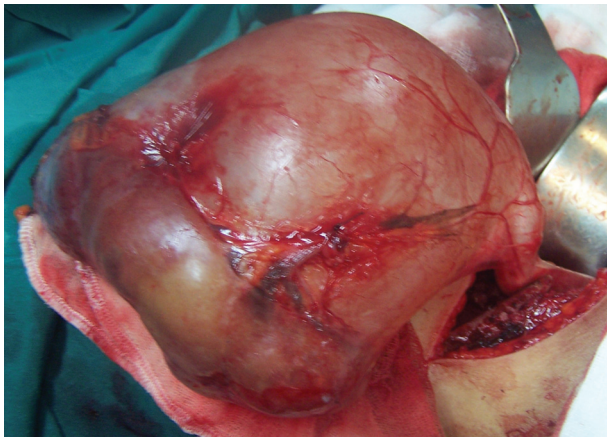

Figure 4. preoperative aspect of GH of the right kidney due to an obstructive ureteral stone

pelvis (n = 6), proximal ureter (n = 1), iliac ureter (n = 4) and distal ureter (n = 2). Stone size ranged from 10 to 32 mm. Retrograde pyelography, performed in patients with renal failure, revealed a GH caused by iliac ureteral stone. GH was secondary to congenital ureteropelvic junction obstruction (UPJO) (n = 11), pelvic stone (n = 6) and ureteral stone (n = 7). Treatment was conservative in 17 patients in whom split renal function was found satisfactory: participation in more than 20% (n = 10) at renal scintigraphy and/or renal parenchyma thickness > 5mm (n = 8) in IVU or CTScan. Treatment consisted in nephrectomy through lumbar incision and subcostal incision (4 and 3 patients respectively) (n = 7), ureteroscopy with pneumatic lithotripsy of ureteral stones (n = 5), pyeloplasty (n = 8), pyelolithotomy (n = 3) and ureterolithotomy (n = 1). Dormia-Basket was used in 3 patients (those with iliac and lumbar urteral stone) to prevent stone push-back in these extremely dilated ureters. A complementary extracorporeal wave lithotripsy (ESWL) was needed to achieve stone free in one case. Of the 11 hydronephrosis secondary to UPJO, 4 had crossed vessels. The quantity of urine drained intraoperatively ranged between 1.1 and 3.5 litres *(Figure 4)*. All patients who underwent surgical treatment had smooth postoperative recovery. No patient developed cardiopulmonary distress during the intervention or post-operatively. Pathologic studies didn't reveal any malignancy. Follow-up was in the range of 9 to 73 months (mean = 32 months) and based on creatinine level, ultrasound and scintigraphy/IVU. Four of the five patients, who presented with high creatinine level, had chronic renal insufficiency and were on dialysis (2 of them had underwent nephrectomy and 2 pyeloplasty).

One patient, who underwent pyeloplasty, developed recurrent pyelonephritis postoperatively. All the 14 patients, who underwent conservative treatment, kept a satisfactory split renal function on repetitive IVP and scintigraphy on follow-up with normal creatinine level. In these patients, latter radiological investigations showed improvement of hydronephrosis in 11 of them. In the three remaining patients, renal dilatation was stable without any signs of aggravation. None of them developed stone recurrence after treatment. Data of patients is presented in *table 1.*

4. Discussion

The definition of GH has been given as the adult renal pelvis containing one litre of urine or 1.6% of body weight (7). Crooks *et al.* has given radiographic criteria for the diagnosis of this condition which included: 1) Kidney occupies a hemi-abdomen, 2) Meets or crosses the midline and 3) about 5 vertebral bodies in length (4). This entity is seen more often in males than in females (2.4:1). More than 500 cases of GH have been reported in the literature (7). GH usually presents as an asymptomatic enlargement of the abdomen noticed by the patient or incidentally by his physician (6, 8). Usually, the patient remains asymptomatic till late stages as the condition progresses slowly (9, 10). He may present with different symptoms like flank pain, hematuria, acute abdomen and recurrent urinary tract infections (9, 10). Moreover, GH kidney may cause intestinal obstruction with constipation and vomiting, respiratory distress, hypertension, venous edema of the lower limbs, obstructive jaundice, and even contralateral ureteropelvic junction obstruction (7, 11). When hematuria is reported, association of urinary stones or cancer must be considered (12).

Since GH is a slowly increasing disease, a large abdominal mass or distended abdomen may be the only sign and this can be very confusing with many cystic abdominal conditions like hepatobiliary cysts, mesenteric cysts, pseudomyxoma, cystic renal tumor, retroperitoneal tumors, ovarian cyst, retroperitoneal haematoma, ascites and splenomegaly (9, 10). Once the presence of GH is known, the possible coexistence of a malignant tumor (especially ureteropelvic tumor) should be considered; thus, it is preferable to conduct a detailed radiological ex-

ploration of the renal pelvis and calyces and to perform repeated cytologic studies of the urine (1). In our cases, no signs indicative of malignancy were found. Diagnosis of this condition is usually done by ultrasonography, excretory, antegrade and retrograde urography. CT scan of the abdomen and magnetic resonance imaging are required for ruling out other differential diagnoses (10).

Plain abdominal radiographs may not always provide an accurate diagnosis due to obscuration of radioopaque urinary calculi by bowel shadows, or overlap with the spine (6). Ultrasound in such patients is a quick, non-invasive and sensitive method for confirming the diagnosis in order to undertake prompt and appropriate management (6). In literature, UPJO was the most common cause of giant hydronephrosis (33%), followed by stones (20%) and congenital ureteral narrowing (7, 10). Other causes include ureteropelvic tumors (2), trauma, renal ectopia (13, 14), retroperitoneal fibrosis (7), obstructive megaureter, ureteric atresia and obstructive ectopic ure¬ter with or without a duplex system (10). All patients with GH do not have similar anatomical configuration and functional status of renal units, and therefore treatment has to be individualized in every patient (10). Usually a strategic approach is followed for treatment of GH. We have formulated ours *(Figure 1)*. Classically a percutaneous nephrostomy is done if the patient is febrile and/or high serum creatinine level or if IVU shows non-visualized unit or pelvicalyceal system is not well delineated (15).

In addition, preoperative percutaneous nephrostomy was proposed to measure unilateral 24-hour creatinine clearance (8), to improve renal function, to prevent severe impairment of renal function, post operative acute renal failure and cardiopulmonary distress. These latter conditions may be due to sudden decompression of the huge hydronephrotic sac, which results in a change in the hemodynamic balance. Therefore, a two-stage procedure with slow decompression by percutaneous nephrostomy before the nephrectomy is preferred (7, 16). This procedure was reported only in paediatric patients and seems not to be so interesting in adults. Thus, it was not performed in our series. On the other hand, function of the GH kidney can be assessed with high sensitivity by pre-operative isotope scanning (8).

Further, based upon overall functional status, ablation of unit or reconstructive surgery is planned (10). The essential aim of treatment of GH should be preservation of the parenchymal loss (9). A kidney participating more than 20% of the total renal function must be conserved. The decision between nephrectomy and kidney-conserving therapy should be a critical issue. However, in practice, nephrectomy has been performed in the majority of instances. Hoffman (17) stated that nephrectomy is often the only therapy for GH because there is no feasible prospect of improvement in renal function especially if the function of the contralateral kidney is normal. The primary reasons are that the existence of the huge hydronephrotic kidney, even when retaining some function, is likely to cause gastrointestinal disturbances from compression and also would be subject to trauma. In addition, the risk of development of malignant tumor due to chronic stimulation by stones, if present, should be considered (1).

Uson *et al.* had a nephrectomy rate of 70%, whereas Crooks *et al.* found it necessary to remove the kidney in only 30% of cases (18). Our nephrectomy rate was 33%. We believe that GH is not an absolute indication for nephrectomy. As reported by Crooks *et al.* (4), the repair is possible in the majority of cases. Pyeloplasty is performed in case of GH due to UPJO. Additionally, calycoureterostomy, calycocystostomy, and Boari flap calycovesicostomy may be indicated in selected cases (6). These procedures are recommended in cases with massive calyceal dilatation and severely compromised peristalsis in the collecting system (19, 20). According to Shah *et al.* (10), patients who had grossly dilated extrarenal pelvis, should undergo reduction pyeloplasty with nephropexy. Nephropexy reduces stasis and improves dependent drainage by tilting the pelvicalyceal system laterally and bringing it more in line with the upper ureter. We believe in our practice that such procedure is not mandatory and doesn't improve the flow of urine. Laparscopy was reported in treating GH (21, 22). In 1999, Hemal *et al.* (23) reported 18 laparoscopic nephrec¬tomies for GH. They performed only 6 cases via transperitoneal approach. In the present series, we performed open nephrectomy in 8 cases because of our limited experience in laparoscopy and also because such nephrectomies seem to be difficult and risky. We, however, performed laparoscopic pyeloplasty in two patients with UPJO.

In the present series, the recovery of renal function and effective drainage following different reconstructive pro¬cedures in different patients substantiate the strategy that anatomical configuration should dictate the type of op¬eration in individual patients.

Careful follow-up, however, will be mandatory (8) to detect any stone recurrence, infection or urinary obstruction. GH in adults is an uncommon clinical entity and often clinically misdiagnosed. Through a series of 24 cases, we have formulated a strategic approach to treat this subset of patients. In a very poorly functioning unit, nephrectomy is the procedure of choice. If the unit is salvageable, type of reconstructive procedure to be selected should be based upon the etiology and location of obstruction.

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Conflict of interest

None declared.

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