Giant Hydronephrosis : Unusual Presentation

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Two cases of giant hydronephrosis are reported and the literatures are briefly reviewed. The first case was unusual in its coincidence of giant hydronephrosis together with full term pregnancy. The second case was the gaint hydronephrosis resulting from calculous occlusion.

The importance of pre-operative diagnosis is emphasized. The chances of infection of the hydronephrotic fluid, the enormous size of the hydronephrosis and the differential diagnosis are discussed.

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A hydronephrosis, containing more than 1000 ml of fluid in children or more than 2000 ml in adults, is generally defined as giant hydronephrosis.

Giant hydronephrosis is a rather rare but a well defined disease entity. Sterling[1] (1939) collected 74 cases of massive hydronephrosis in a review of the world literatures and coined the term "Giant Hydronephrosis". Campbell and associates[2] increased the number to 100 cases in a review up to 1950 and reported on one case. Tombari and associates[3] found 60 additional reported cases during 1950 to 1968 and presented one of the largest case of hydronephrosis since the case recorded by Glass[4] in 1746. Every previous reports presented not more than two cases and almost all of them had single case experience. About 163 cases so far have been reported in the literatures.

The average volume in majority of cases are from 2000-8000 ml. The following is a list of the massive quantity of giant hydronephrosis referred to by Dennehy[5] ·

Earlam (1950)	18,000 ml.
Franck (2 cases)	30,000 ml.
Javal and Dumont	30,000 ml.
Dumreicher	36,000 ml.
Tombari and et al. (1968)	52,000 ml.
Glass (1746 - autopsy)	115,000 ml.

The gaint hydroneprosis is mostly associated with non-functioning and atrophy of the renal parenchyma. It is most important to make a correct initial diagnosis. Gaint hydronephrosis was often clinically mistaken for ascites, which frequently led to paracentesis with the subsequent development of renal infection.[2,3,5] Acute pyelonephritis and septic shock following the tapping of hydronephrotic kidneys have been documented. Pre-operative erroneous diagnosis of the giant hydronephrosis comprised 54 per cent (Table 1). This resulted in a transperitoneal procedure in stead of the retro-peritoneal approach in many cases.

Table 1 Categorically list of pre-operative erroneous diagnosis.

Ascites
Retroperitoneal cyst or tumour
Abdominal mass
Pancreatic cyst
Hypernephroma
Mesenteric cyst, Choledochus cyst, Hydatid cyst, Adrenal rest tumour, Splenic tumour, Neurofibroma, Pregnancy, Acute urinary retention.

The congenital obstruction at the pelvi-ureteric junction is the most common cause (87 per cent) of giant hydronephrosis.[2,5] Traumatic ureteric stricture accounts for 12 percent in producing giant hydronephrosis.[2] Only one case reported by Wyrens was preceded by calculous obstruction. (Table 2).

Table 2 The etiologic factors in giant hydronephrosis.

1. Congenital obstruction-most common (87%), mainly at pelvi-ureteric junction
2. Traumatic ureteric stenosis (12%), around upper ureter or at pelvi-ureteric Junction
3. Calculous obstruction (only one case).

One stage excision of giant hydronephrosis had been utilized in most cases reported previously.

Tombari et-al.[3] (1968) excised a 52 litres hydronephrosis from a 32 year-old man, the patient's vital signs remained stable through out the procedure and the convalescence was uneventful. Holle[7] (1976) stressed the hazards in the management of large intra-abdominal masses, the consequences of sudden decompression, leading to cardio-respiratory distress and this may threaten life. Talukder[8] (1979) suggested two stage procedure for an extensive and massive hydronephrosis, slow decompression by tapping initially and a definitive surgical removal of the hydronephrotic sac 2 weeks later.

In the Division of Urology, Rajvithi Hospital, Bangkok from 1968 to 1974, the records of 235 cases hydronephrotic kidney were analysed.[9] Only 2 cases fell into the category of the giant type and are worthy of a detailed report.

The first patient carried on with her first pregnancy until delivery without any distressing symptom from the large hydronephrotic kidney. In addition, the compression produced by this huge hydronephrotic sac on its renal pedicle resulting in complete obliteration of the renal vessels observed at the operation. Thus, congenital absence of the kidney suggested preoperatively by selective renal angiogram was erroneous.

The second case report in this paper is considered the second case of the giant hydronephrosis caused by calculous obstruction since Wyrens reported his first case in 1949.

CASE REPORTS

CASE 1 A 22-year-old female was admitted to Rajvithi Hospital for the delivery of her first baby.

On examination of the abdomen, a large mass was palpated and was considered to be a right ovarian cyst displacing the full-term pregnant uterus to the left. Such displacement by soft tissue mass was confirmed by a plain x-ray of the abdomen. (Figure1).

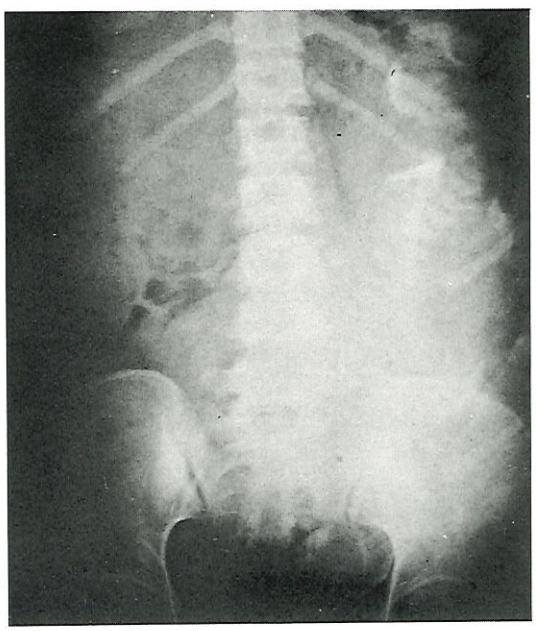

Fig. 1. Plain abdomen : Full term pregnant uterus displaced by large soft tissue mass to the left.

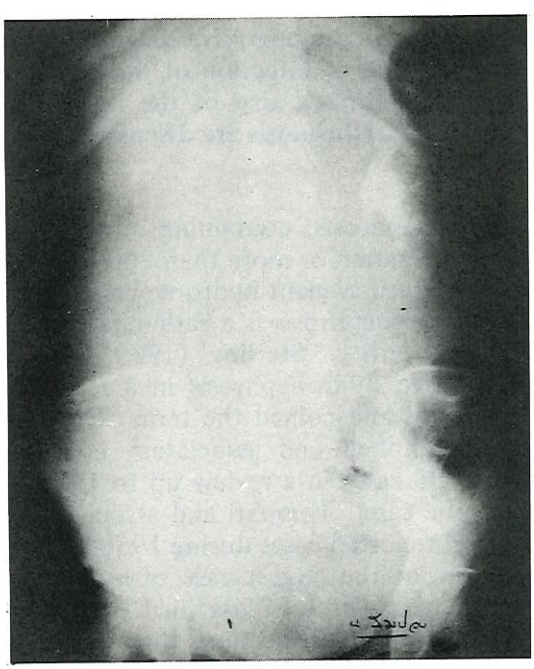

Fig. 2. Excretory urogram. Non visualized right kidney, normal left kidney with deviation of left ureter laterally. Soft tissue mass on the left of the abdomen was noted.

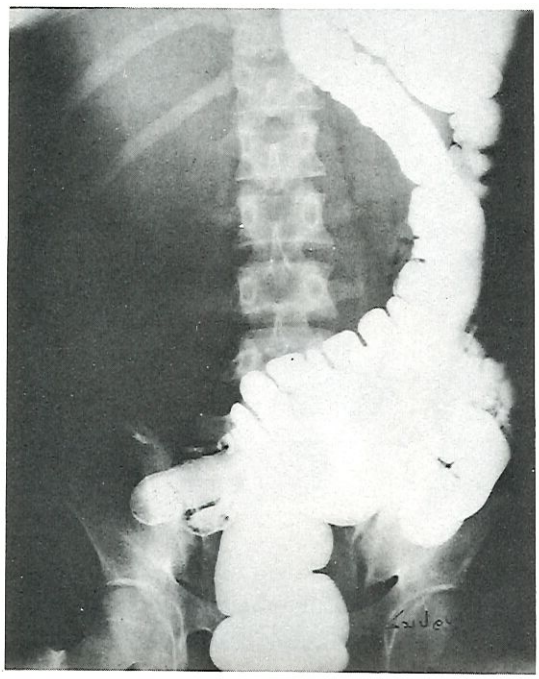

Fig. 3. Barium enema in case 1.

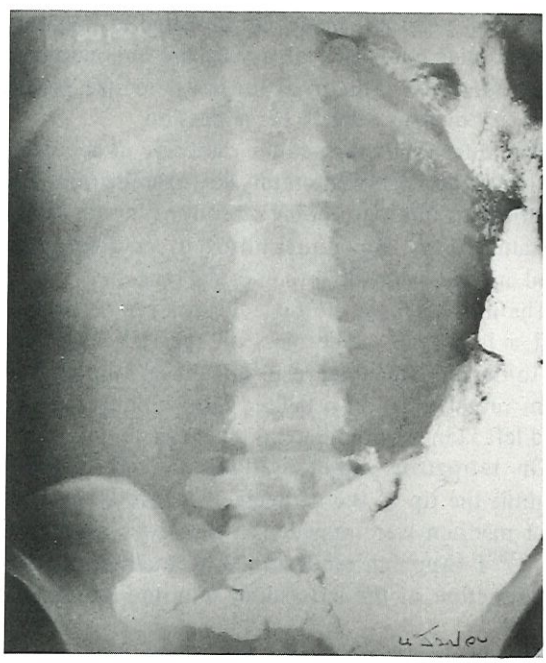

Fig. 4. Upper GI and small bowel study in case 1.

Additional history was given by the patient that she had noticed an abdominal mass since the age of 7 and the mass had increased in size gradually but with very little discomfort.

Excretory urogram, after the delivery of a full term baby, revealed a normal functioning hypertrophic left kidney with deviation of left ureter laterally, non-visualization of the right kidney and a soft tissue mass on the right of the abdomen. (Figure 2).

Polycystic kidney was suspected and the patient was transferred to the care of urologist.

Barium enema, upper GI and small bowel series were performed respectively. A huge soft tissue mass in the right retroperitoneal space displacing stomach, intestine and right half of the colon to the left was radiologically demonstrated (Figures 3, 4).

Left retrograde pyelogram appeared normal. The right retrograde catheter was impeded in the lower half of the ureter and no contrast medium filled either the upper ureter or the pyelocalyceal system. The right catheter was seen curving to the left lower border of the soft tissue mass (Figure 5).

The renal angiogram suggested congenital absence of the right kidney and confirmed a huge retroperitoneal mass on the right side (Figure 6).

Exploratory laparotomy through long right paramedian incision disclosed a large thin wall hydronephrosis of the right kidney. Atrophic obliteration of the renal vessels was also noted (Figure 7). The hydronephrotic kidney was removed with incidental excision of an accessory spleen and appendectomy. The paper-thin hydronephrotic sac containing 6000 ml of clear yellow fluid, was dissected and this disclosed a stenosis at the pelvi-ureteric junction (Figure 8). Analysis of hydronephrotic fluid revealed BUN 60 mg%, creatinine 22.6 mg%, uric acid 12.7 mg%, sugar 65.7 mg%, sodium 110 mEq/l, potassium 10 mEq/l, chloride 87 mEq/l and calcium 8.7 mg%.

The post operative course was uneventful. Barium enema on the twentieth postoperative day showed that the ascending colon and the small intestine had returned to their original position (Figure 9).

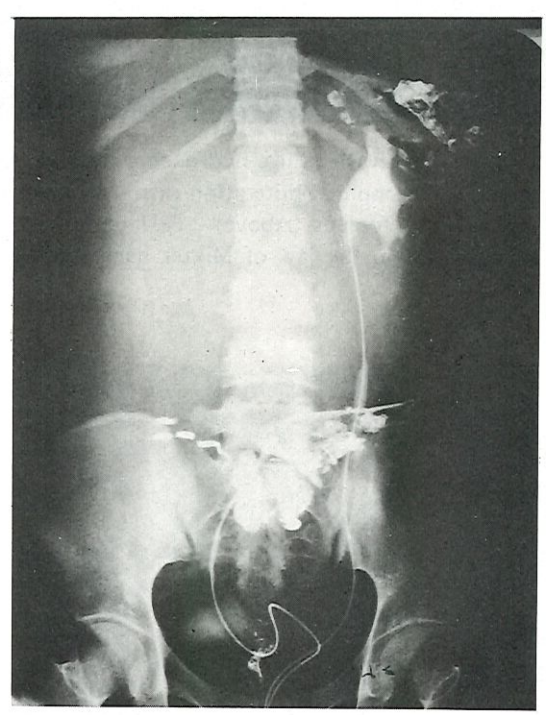

Fig. 5. Retrograde uretero-pyelogram : The right retrograde catheter seen curving to the left of the soft tissue mass. Noted barium in the colon (case 1).

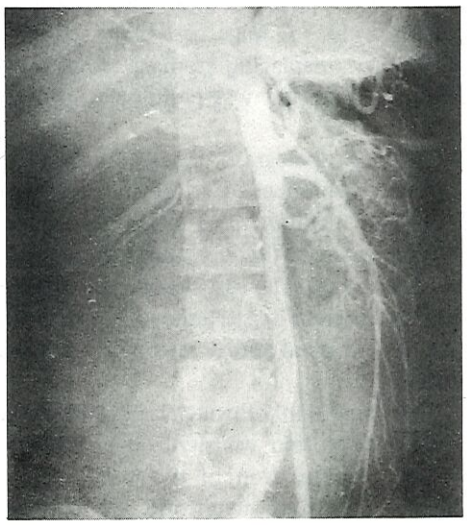

Fig. 6. Selective renal angiogram suggesting congenital absence of right kidney in case 1.

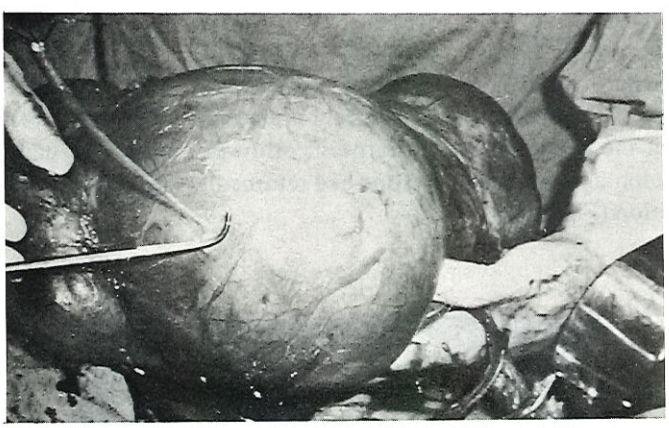

Fig. 7. Exploration revealed huge hydronephrotic right kidney (case 1). Atrophic obliteration of renal vessels in the jaw of Pean forceps (Above). Pelvi-ureteric junction was pointed in the jaw of Mixter hemostatic forceps (Below).

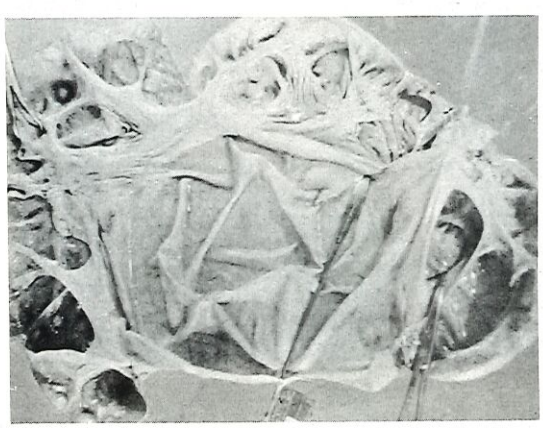

Fig. 8. Paper-thin hydronephrotic sac of case 1. Pelvi-ureteric junction stenosis at the end-of probe.

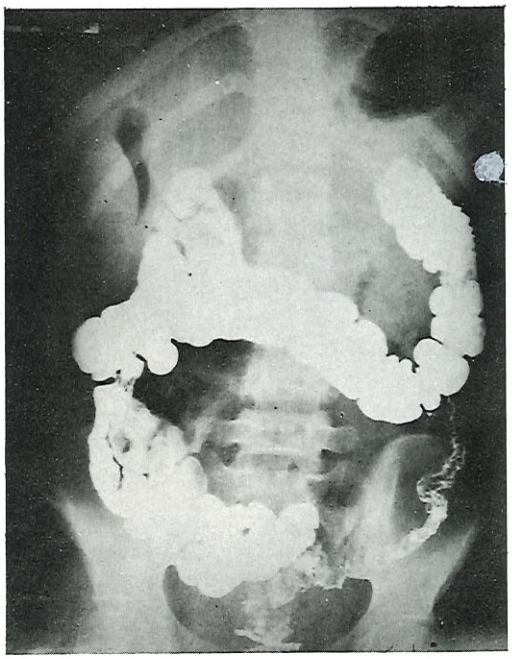

Fig. 9. Barium enema on the 20th P.O. day (case 1).

CASE 2 A 64 year-old female was admitted to Rajvithi Hospital with a chief complaint of a palpable abdominal mass of one-year duration. This mass increased in size rapidly over the last three months prior to her admission.

The tense cystic mass in the right side of abdomen crossing to the left was palpated on physical examination. There was no significant abnormal finding on routine blood, urinary, and stool examinations. Blood urea nitrogen, creatinine, sugar, protein and amylase were all in the normal range.

The liver scan was normal. Plain x-ray of the abdomen showed a large soft tissue mass on the right side extending across to the left. Bilateral renal calculi were noted. Excretory urogram revealed a non-visualised right kidney and poorly-visualised left kidney with bilateral renal calculi.

On retrograde catheterisation, the right ureter would only admit the tip of the catheter. The ureteric catheter and contrast medium were impeded in the lower third of the left ureter. ^{131}I Hippuran renal scan was obtained and showed a normal function of the left kidney and non-functioning right kidney.

The upper GI and small bowel study showed a huge intraperitoneal mass displacing the stomach and the intestines to the left, upwards and downwards.

A diagnosis of a huge hydronephrotic right kidney and bilateral renal calculi was made.

Exploration of the abdomen through a long midline incision demonstrated a large lobulated hydronephrosis of the right kidney. An obstructing stone in the mid ureter was disclosed. Nephrectomy was carried out together with its upper two third of the ureter (Figure 10, 11). Convalescence was unremarkable.

The gross specimen of hydronephrotic kidney was 4.5 kg in weight containing 5000 ml of clear brown fluid (Figure 12).

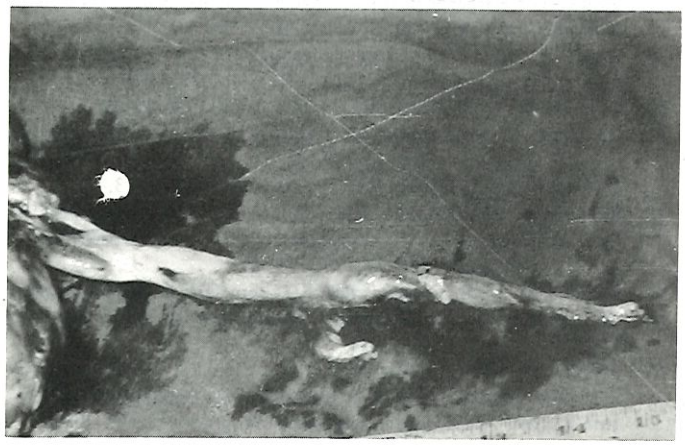

Fig. 10. Obstructing stone seen in the middle of ureter (case 2).

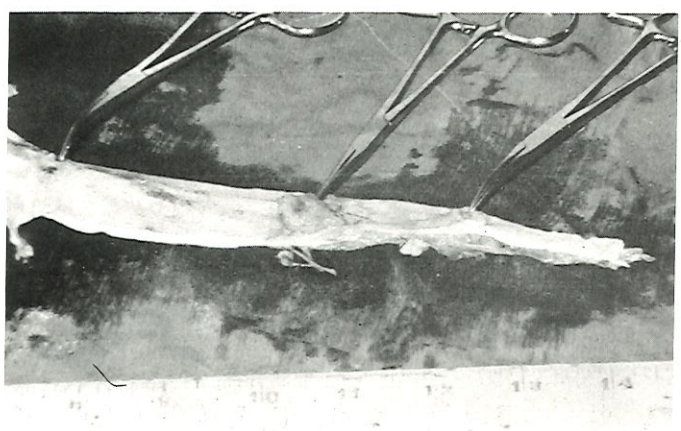

Fig. 11. Obstructing stone pointed by straight hemostat (case 2).

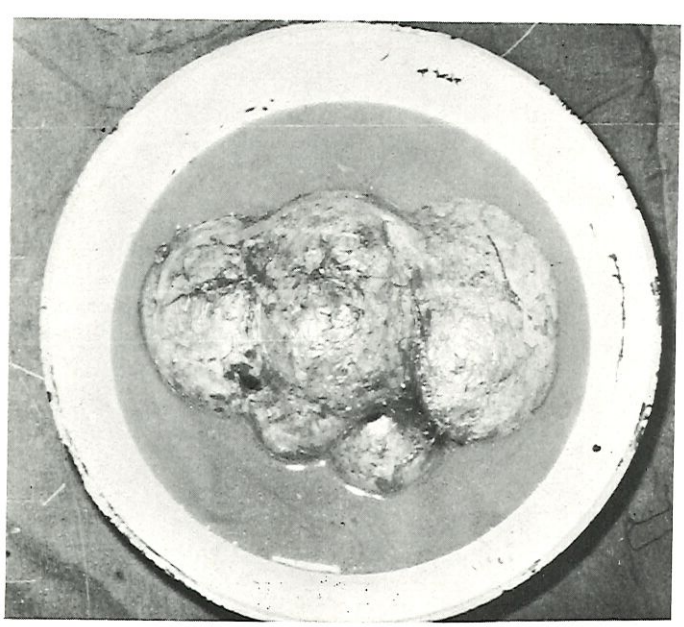

Fig. 12. Hydronephrotic kidney 4.5 kg in weight (case 2).

Culture of the fluid content revealed no bacterial growth. Marked atrophic renal parenchyma and chronic pyelonephritis were reported microscopically.

DISCUSSION

The significance of the giant hydronephrosis lies in the correct diagnosis made prior to surgery. Although complete investigation on our first case was carried out; the appropriate impression could not be achieved. In the second case, with the former experience the diagnosis of huge hydronephrosis was made earlier during the investigation.

In the differential diagnosis of giant hydronephrosis, one must keep in mind this entity and to rule out ascites, retroperitoneal or intra-peritoneal tumour and cyst. Renal scan using technetium per technetate diethylene triamine penta-acetic acid (99m. Tc DTPA) recommended by Raynaud and associates [10] may be helpful in detecting functionless hydronephrosis and to delineate the exact size and limit of the sac. In our first reported case in which the renal vessels were completely obliterated the use of 99m. Tc DTPA imaging may pose a problem. Recently, grey scale ultrasound used in detecting hydronephrosis has been investigated by Ellenbogen et al with encouraging results.[11] Computed tomography may also provide the diagnosis.[12,13]

Stenosis at the pelvi-ureteric junction in the first case is thought to be the result of congenital malformation. Undoubtedly in the second case, the calculous obstruction is the etiologic factor.

In spite of the long-standing hydronephrosis in the first case (approximately 20 years duration), the fluid content in the hydronephrotic sac appeared clear and clean indicating very little, if any infection. This is a similar feature in a great number of cases reported in the literatures. The retardation of infection has not been explained clearly, possible reason may be that stricture at the pelvi-ureteric junction hinders the ascending infection. Surprisingly enough, the hydronephrotic fluid of our second reported case was sterile. The brown colour of the fluid content may represent some old hemorrhage of the renal parenchyma during the course of the disease.

The enormous capacity of the hydronephrosis is noteworthy. The etiologic factors producing the great amount of hydronephrotic fluid has been discussed in the texts. There seems to be no doubt that some intermittent blockage at the pelvi-ureteric junction, what ever the cause may be, is said to be the prime factor. Extra-renal type of renal pelvis tends to be markedly dilated. As a general rule, the rate and degree of hydronephrosis depends on the renal parenchyma-the nephrons and the result of imbalance between formation, resorption and excretion of the urine.

REFERENCES

1. Sterling, WC : J Urol 42:520, 1939. Cited by Campbell, EW, et al.
2. Campbell, EW, Hunsicker, WC and Marconis, J : Giant Hydronephrosis of Traumatic Origin, A Case Report. J Urol 63:970, 1950.
3. Tombari, AA, Power, RF, Harper, JM et al : Giant Hydronephrosis, A Case Report with Review of Literature. J Urol 100:120, 1968.
4. Glass, S : Cited by Dennehy PJ
5. Dennehy PJ : Giant Hydronephrosis in a Double Kidney. Br J Urol 25:247, 1953.
6. Wyrens, RG : J Urol 61:311, 1949. Cited by Campbell EW, et al.
7. Hoile RW : Harzards in the Management of Large Intraabdominal Tumours. Ann Roy Coll Surg 58:393, 1976.
8. Talukder, BC : Two Stage Procedure for the Management of Massive Hydronephrosis. J Urol 122:391-393, 1979.
9. Sangmit, S : Hydronephrosis. J Urol Thai 1:81, 1976.
10. Raynaud, C, Koutoulidis, C, Cendron, J et al : Pre-operative Identification of a Hydronephrotic Sac with Sequential ^{99m}Tc DTPA Imaging : Case Report. J Urol 109:1033, 1973.
11. Ellenbogen, PH, Scheible, FW, Talner, LB, et al. : Sensitivity of Grey Scale Ultrasound in Detecting Urinary Tract Obstruction : Am J Roentgenol 130:731-733, 1978.
12. Williamson, Jr. B, Hattery, RR, Stephen, DH, et. al : Computed Tomography of the Kidneys : Semin Roentgenol 8:249-256, 1978.
13. Forbes, W. ST. C, Ischerwood, I, Fawcitt, RA : Computed Tomography in the Evaluation of the Solitary or Unilateral Non Function Kidney : J Computer Assisted Tomography 2:389, 1978.