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# Megacalycosis or Puigvert Disease, a Rare Congenital Calyceal Anomaly: A Report of 3 Cases

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**ABSTRACT**

Megacalycosis is most likely congenital. It is primarily a disease of the renal calyces, which is usually diagnosed because of its complications, such as calculi or infections in the urinary tract. It has also been diagnosed prenatally. In the absence of complications, the disease is discovered accidentally in the course of urologic examination undertaken for a different reason. This article reports 3 cases of megacalycosis presenting to us with varying symptoms. Awareness of the possible existence of megacalycosis in patients suffering from infections and calculi in the urinary tract contributes to and facilitates the choice of appropriate treatment.

## INTRODUCTION

Congenital megacalycosis is a rare renal disorder consisting of calyceal dilatation without evidence of renal, pelvic, or ureteral obstruction. The following criteria should be met: The calyces should be dilated, the kidney must be large with polycalycosis, the infundibulum, pelvis, and ureter should have normal calibers and isotopes, and the renogram should show normal renal function with no outflow obstruction. Since its initial description in 1963, only a few cases have been reported. The defect is mostly bilateral, showing male predominance, and it does not disrupt the renal functions. We report on 3 male patients, 2 with unilateral megacalycosis and 1 with bilateral megacalycosis, and different clinical presentations. It is important to bear in mind the possibility of megacalycosis in the differential diagnosis of hydronephrosis to prevent unnecessary diagnostic and therapeutic intervention.

## CASE REPORT

Our first case, a 32-year-old male, presented with a urinary tract infection (UTI) and bilateral renal cysts. The physical examination was unremarkable. Biochemical tests of renal function were normal, except for a positive urinary culture treated accordingly. The renal ultrasonogram displayed enlarged kidneys and caliectasis with normal corticomedullary

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differentiation. The intravenous urography showed dilated calyces on both sides (Figure 1), with a normal pelvis and ureter. The isotopic renogram showed no outflow obstruction. The patient followed up with an annual IVU, with similar anatomical pictures.

Figure 1. IVU showing bilateral megacalycosis.
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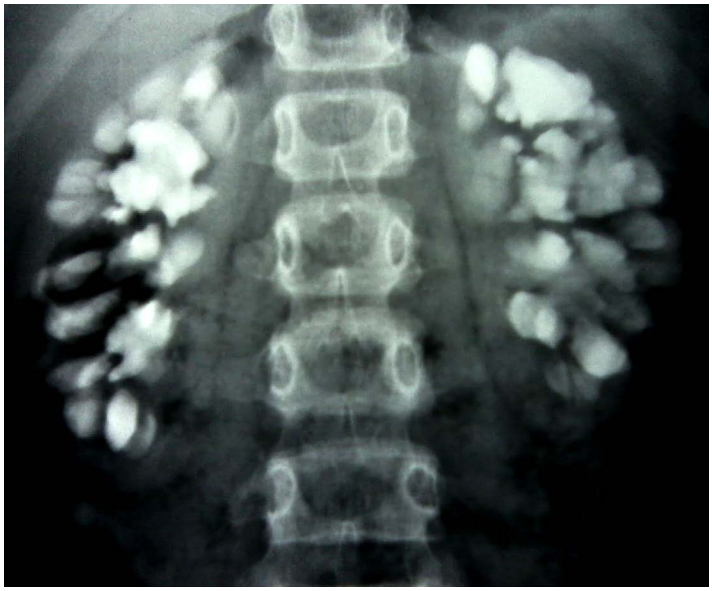

Our second case, a 40-year-old male, presented with pain in the right hypochondrium with suspicion of having gallstones, but the ultrasound (US) showed left-sided hydronephrosis. No other symptoms were present. The physical examination did not reveal any positive findings. On further evaluation with IVU, he was diagnosed with a case of left-sided megacalycosis (Figure 2). Upon regular follow-up, he demonstrated normal renal function, and no outflow obstruction on the isotope renogram was seen.

The third case, a 45-year-old male, had mild, dull aching pain over his left flank without any radiation. The physical examination was unremarkable. On ultrasonography (USG), there was an enlarged, left-sided kidney and mild pelvicalyceal dilatation with normal corticomedullary differentiation. The IVP showed left-sided, faceted calyces with polycalycosis (Figure 3), and a normal pelvis and ureter. The diuretic renogram ruled out any outflow obstruction. The patient was put on antispasmodics and is on regular follow-up without any symptoms.

All the cases have normal urinalysis, normal hematological and biochemical tests, and sterile urine cultures.

## DISCUSSION

Megacalycosis is best defined as the nonobstructive enlargement of calyces resulting from malformation of the renal papillae. There is frequent bilateral involvement, but unilateral cases are common. It is a rare condition first described by Puigvert in 1963 [1]. The pathogenesis has not been clearly established [2]. It predominantly occurs in males with a ratio of 6:1. It was theorized by Puigvert (1964) and endorsed by Johnston and Sandomirsky (1972). There is a transient delay in the recanalization of the upper ureter after the branches of the ureteral bud hook up with the metanephric blastema. This produces a short-lived episode of obstruction when the embryonic glomeruli start producing urine. The fetal calyces may dilate and then retain their obstructed appearance despite the lack of evidence of obstruction in postnatal life, characterized by enlarged kidneys and an occasionally outnumbered, uniform dilatation of calyces (polycalycosis). The number of calyces usually increases from 8 to 10 to as many as 20 to 25. Renal function and radiographic appearances have remained stable in cases followed over several years, demonstrating a mild distal tubular concentrating defect [3]. Infudibula, pelvis, and ureter are usually normal. The cortical tissue around the abnormal calyx is normal in thickness and shows no signs of scarring or chronic inflammation. The medulla, however, is underdeveloped and assumes a falciform crescent instead of its normal pyramidal shape. The collecting tubules are not dilated but are definitely shorter than normal, and they are oriented transversely rather than vertically from the corticomedullary junction. Megacalycosis in the duplex upper moiety have been reported [4]. The disease is usually diagnosed because of its complications (renal calculi and UTIs) and calyceal stasis, or it is discovered accidentally [3]. The diagnosis lies in the typical radiographic findings of the IVU and renal ultrasonogram, coupled with the normal washout pattern in renal scintigraphy [4,5]. In IVU, the calyces appear faceted, the kidney is large with multiple calyces, and the infundibula, the pelvis, and the ureter should have a normal caliber. All the cases are under close follow-up, focusing on the prevention of urinary tract infections. Urolithiasis with active urological

Figure 2. IVU showing left-sided megacalycosis.
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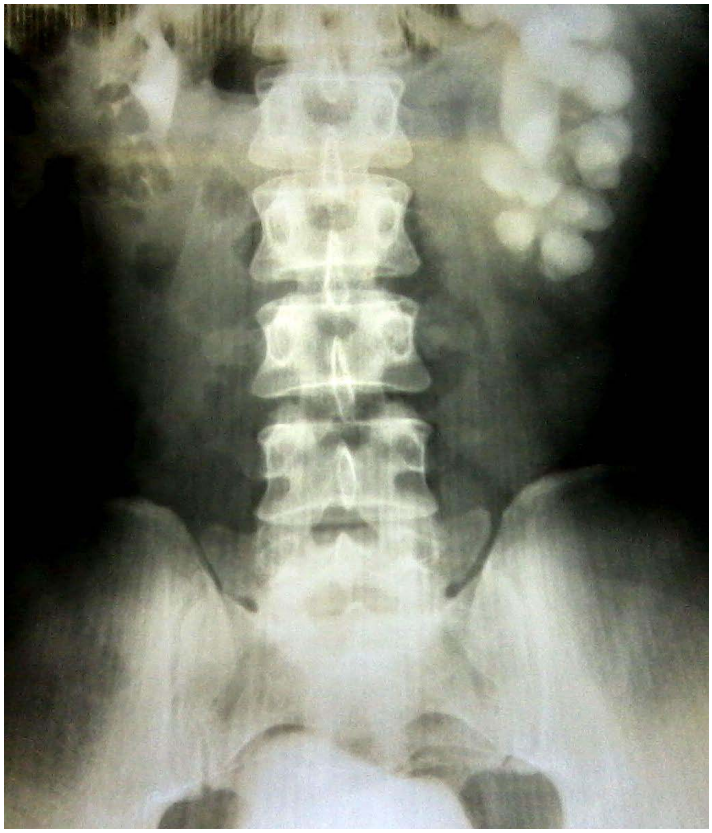

Figure 3. IVU showing left-sided megacalycosis.
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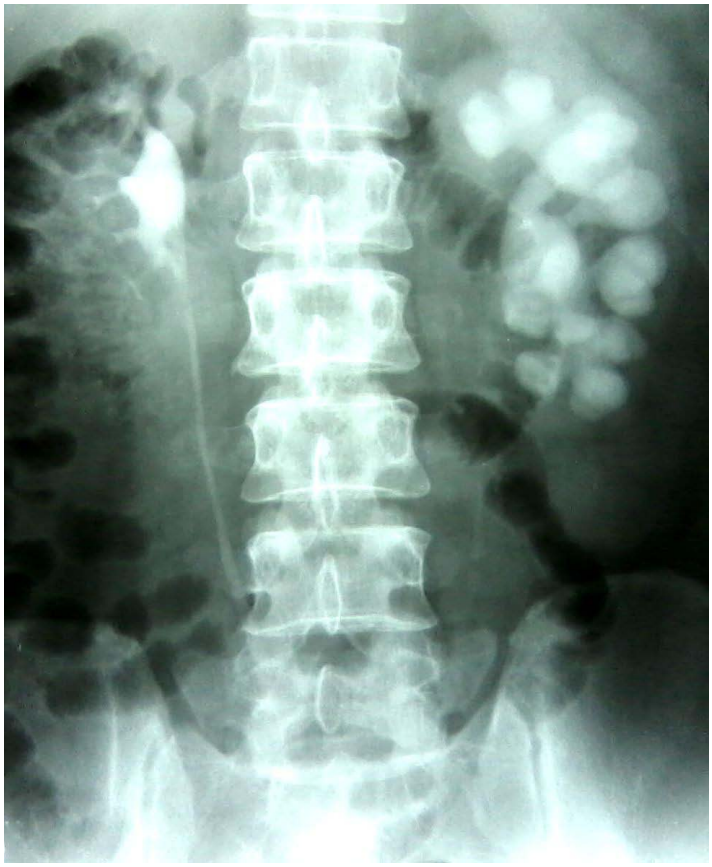

intervention is not required in any case, which is similar to the management in other case reports.

We believe retrograde pyelography and micturating cystourethrography should be reserved, as these are invasive procedures and may introduce infection into the urinary tract and add to the cost. Megacalycosis should be considered in the differential diagnosis of calyceal dilatation to prevent unnecessary therapeutic intervention. An annual IVU should not reveal any progression of the anatomic derangement or functional impairment of the kidney. Intervention is required only when secondary complications like nephrolithiasis and infections occur. As this is a rare clinical entity, the number of cases is small, which is similar to other case reports.

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