GRADE IV VESICOURETERAL REFLUX ON SINGLE FUNCTIONING KIDNEY OR PENTALOGY OF FALLOT: WHICH CONDITION HAS TO BE CORRECTED FIRST? A CASE REPORT

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Abstract: *Tetralogy of Fallot is a congenital heart disorder which comprises right ventricular outflow tract obstruction, ventricular septal defect, aorta dextroposition and right ventricle hypertrophy. Complicated with an atrial septal defect or patent ductus arteriosus, it is called pentalogy of Fallot. Vesicoureteral reflux (VUR) is characterized by retrograde flow of urine from the bladder to the kidneys, and may be associated with urinary tract infection, hydronephrosis and abnormal kidney development (renal dysplasia). Both conditions above have a poor influence on renal function. The order and the method of the treatment represent a challenge when they affect the same patient. Case report: A 14 month-old male, born in a territorial hospital in Romania, was sent to our service for clinical evaluation and possible surgical correction. By the age of 5 months, pentalogy of Fallot and VUR grade IV on the single functioning kidney was diagnosed. After proper preoperative investigations, right nephroureterectomy and Cohen transvesical left ureteal reimplantation were made in our service. Postoperatively, after 25 hours, the patient died in the intensive care unit, the determined cause of death was sudden cardiac arrest. Conclusions: Surgical correction sequence is problematic, as each condition has serious influences on the other. An important question emerges: which defect has to be corrected first: VUR grade IV on single functioning kidney or pentalogy of Fallot?*

INTRODUCTION

Tetralogy of Fallot (TF) is one of the most common congenital heart disorders and comprise right ventricular outflow tract obstruction (infundibular stenosis), ventricular septal defect, aorta dextroposition, and right ventricle hypertrophy. If it is complicated with an atrial septal defect (ASD) or patent ductus arteriosus, it is called pentalogy of Fallot (PF). The condition is classified as a cyanotic heart disease because of the right-to-left shunt. The ideal management of this pathology is primary repair, performed around the age of 12 months, which has excellent short-term outcome. Postoperative morbidity includes arrhythmias, postoperative bleeding, temporary renal failure, and neurologic injury.(1,2,3,4)

Vesicoureteral reflux (VUR) is characterized by the retrograde flow of urine from the bladder to the kidneys, and may be associated with urinary tract infection (UTI), hydronephrosis, and abnormal kidney development (renal dysplasia). Unrecognized VUR and accompanying urinary tract infection increases risk of pyelonephritis, hypertension and progressive renal failure. The management of this condition is based on literature reviews, which highlight that spontaneous resolution of vesicoureteral reflux is common in young children but is less common as puberty approaches. Sterile reflux does not result in reflux nephropathy and long-term antibiotic prophylaxis in children is safe. Surgical treatment is reserved for patients presenting breakthrough febrile UTIs despite adequate antibiotic prophylaxis, severe reflux (grade V or bilateral grade IV), mild or moderate reflux in females that persists as the patient approaches puberty, poor compliance with medications or surveillance programs and poor renal growth or function or appearance of new scars, and it is highly successful in experienced hands.(5)

Both conditions above have a poor influence on renal function. The order and the method of the treatment represent a challenge when they affect the same patient. We present a case of a 14 month-old boy admitted in our service suffering from PF, VUR stage IV on single functioning kidney, chronic renal failure, and right kidney hypoplasia.

CASE PRESENTATION

A 14 month-old male from a physiological birth, followed properly prenatally by the family doctor and the obstetrician, vaginal birth, with 3770 grams of weight, APGAR score 9 at 1 minute, vaccinated according to WHO schedule excepting BCG. He was directed by a pediatric nephrologist to our service. Prenatally, the mother had not been informed about any kind of malformations. The patient was born in a territorial hospital. At the age of 2 months, because of episodes of bluish pale skin during crying and feeding, cyanosis of the lips and nail bed, a cardiac malformation was suspected, but only at the age of 8 months, PF was diagnosed. The patient was directed to a cardiovascular surgeon. In meantime, at the age of 5 months, the child presented an episode of fever, vomiting, anorexia and lethargy. A UTI with E. Coli was diagnosed. He was presented to Pediatric Nephrology Department for a consult and renal function evaluation. On CBC, leukocytosis and neutrophilia were highlighted, normal ranges in urinalysis and biochemical analysis were obtained, and E Coli was found in the urine culture. Estimate glomerular filtration rate (creatinine clearance) was elevated, using the Schwartz method and revised Schwartz method. Right kidney dystrophy and grade 2 left hydronephrosis

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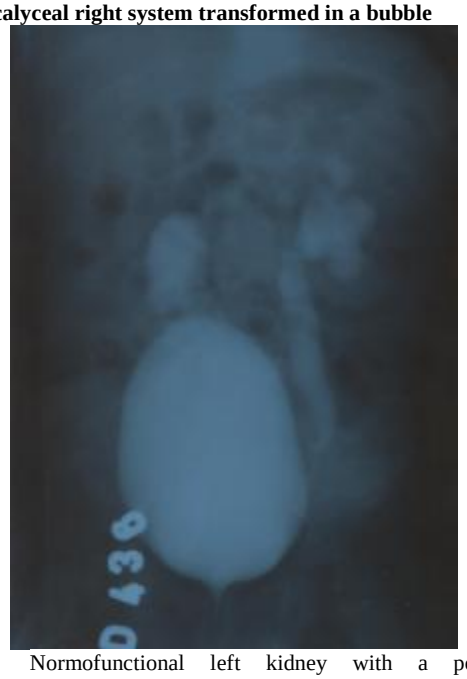
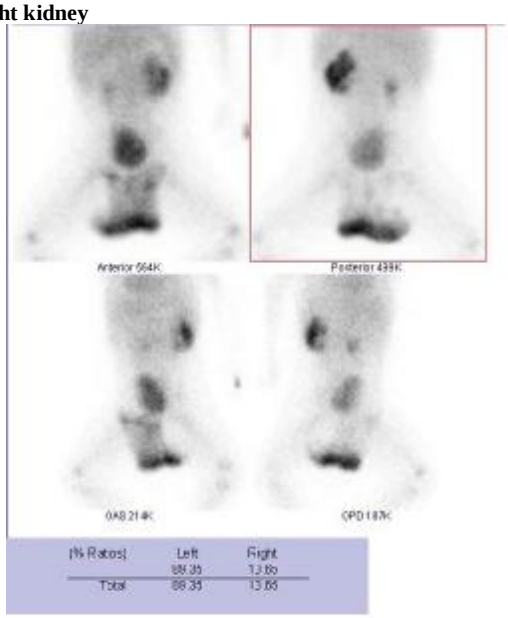
were found with an abdominal ultrasound. Renal secretion and excretion delayed on the right side, disorganized bubble-like right pyelocalyceal system and IIIrd-IVth degree left hydronephrosis were emphasized with intravenous urography. Grade III-IV bilateral VUR was obtained after voiding cystourethrogram (VCUG) with Iomeron (figure no. 1).

Figure no. 1. VCUG-bilateral grade III-IV VUR and pelvocalyceal right system transformed in a bubble



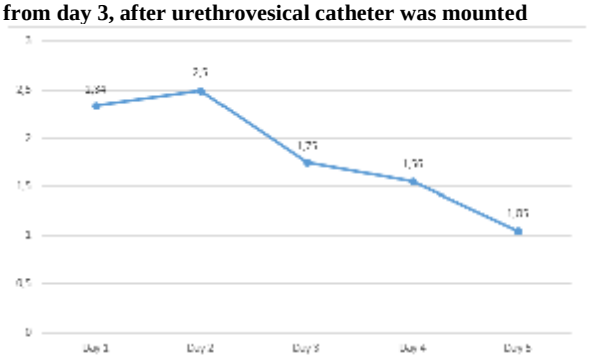
Normofunctional left kidney with a possible malrotation and hypoplastic hypofunctional right kidney, with a differentiated function LK/RK of 89.35%/10.65% were found on Tc DMSA scan (figure no. 2).

Figure no. 2. Tc DMSA scan- Normofunctional left kidney with a possible malrotation and hypoplastic hypofunctional right kidney



Antibiotic treatment was administrated during hospitalization. The patient was discharged on chronic chemoprophylaxis in order to be evaluated from cardiovascular viewpoint. Fallot tetralogy with pulmonary vascular ring hypoplasia and ostium secundum atrial septal defect were found after cardiac evaluation. Surgical correction was recommended to be delayed until renal status is balanced. A new nephrological evaluation after 5 months found a degradation in renal condition, and urological assessment was recommended. Taking into account the evolution of the case, surgical treatment was considered imminent. The patient was hospitalized in our clinic in relatively good general condition. Biochemical evaluation indicated high levels of urea and creatinine, but after an urethrovesical catheter was mounted, the decrease of these values was noticed (figure no. 3). Posterior urethral valve was excluded by a voiding cystography. A balanced cardiovascular function was found in preoperative cardiac evaluation, but because of the renal status, the cardiac surgical correction was contraindicated. He was admitted to surgery and underwent right nephroureterectomy and Cohen transvesical left ureteral reimplantation. Postoperatively, the patient was transferred in ICU, where he had a good progress, he was stable hemodynamically and respiratory and he had normal fluid balance and normochromic urine on bladder drainage after 6 hours. After 25 hours, without any prodrome, the patient suffered a cardiopulmonary arrest unresponsive to resuscitation. No major organic lesions were highlighted by autopsy, which concluded a sudden cardiac arrest.

Figure no. 3. The evolution of creatinine levels during first 5 days of hospitalization. We notice that the values are decreasing towards normal values (0.72-1.25 mg/dl) starting from day 3, after urethrovesical catheter was mounted



DISCUSSIONS

Acute kidney injury is an important prognostic complication of cardiac surgery. Many studies have shown a high prevalence of this complication. Most of them estimated that more than 30% of cardiac surgery patients develop clinically relevant kidney injury.(6) It remains an important cause of morbidity and mortality. The etiology of renal insufficiency following cardiac surgery is poorly understood but it is believed that it results from inadequate perfusion, exotoxins and endotoxins. Some of the risk factors of acute kidney injury were identified by previous studies. Preexistent kidney disease, as in our patient, is one of them, and it is presented as non-modifiable.(7)

The management of vesicoureteral reflux has been a matter of debate for decades. Surgical options for correcting VUR are endoscopic, open, laparoscopic and robotic procedures. In higher grades of VUR (III, IV, and V), there is no definite advantage of medical management over surgery.(8) Endoscopic hyaluronic acid/dextranomer injection is a

minimally invasive procedure, which requires general anesthesia and it can be done as a day care procedure. In Grade IV VUR, 1st injections resolved VUR in 83.3% ureters. The studies report comparable results in higher grades of VUR (94% resolution rate after three sessions of injection hyaluronic acid/dextranomer) to the open ureteric reimplantation (95% resolution rate) after 2nd and 3rd injection.(9,10) Open repairs prevent reflux by increasing the length of the intravesical ureter, facilitating compression of the ureter against the detrusor muscle during bladder filling. Laparoscopic and robotic surgery demonstrated no significant improvement in outcomes compared to open surgery, excepting hospitalization time and postoperative analgesic treatment, but the length of surgery and anesthetic risks are higher.(11) The estimated success rate of open surgery is 98.1% (95% CI 95.1, 99.1).(12) Various open approaches have been described including extravesical, intravesical, as well as combined approaches. The technique currently used and indicates by most specialists is described by Cohen in 1975. The ureter is advanced through a submucosal tunnel across the trigone to the contralateral bladder wall with the new mucosal orifice located superior to the contralateral orifice.(13,14)

Decisions for surgical repair and type of repair are individualized for VUR. Considering our patient status and comorbidities, we aimed to choose a surgical option with maximum efficiency and surgical and anesthetic risks as low as possible so we used Cohen technique for antireflux ureteral reimplantation.

CONCLUSIONS

The management of high grade VUR on a single functional kidney is difficult when the patient has such important cardiac defects as pentalogy of Fallot. Surgical correction sequence is problematic, as each condition has serious influences on the other. An important question emerges: which defect has to be corrected first: VUR grade IV on single functioning kidney or PF?

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