

Trauma and reconstruction

## “Laparoscopic pyeloplasty for treating ureteropelvic junction obstruction in a patient with Fontan physiology: The first case described in literature”

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## ABSTRACT

The Fontan procedure is a palliative operation for patients with single ventricle congenital heart disease. In this case-report, we present the case of a 36-years-old man, with Fontan physiology and ureteropelvic junction obstruction, undergoing laparoscopic pyeloplasty. He presented with right flank pain and mild hydronephrosis of his right kidney. Although few laparoscopic operations have been described in the literature, this, to our knowledge, is the first laparoscopic urological procedure described in a patient with Fontan physiology. Laparoscopic pyeloplasty in patients with Fontan physiology, is an efficient and safe technique when performed in centers with extensive experience in laparoscopic procedures.

## 1. Introduction

The Fontan procedure is a palliative operation, performed for patients suffering from single ventricle congenital heart disease. It leads the systemic venous return directly into the pulmonary arteries, resulting in passive filling of the pulmonary circulation without the need of a ventricular contraction. This modification prevents intracardiac mixture of arterial and venous blood, leading to increased oxygen saturation of arterial blood and protecting the single ventricle from volume overload (Fig. 1).<sup>1</sup> Nevertheless, the pulmonary impedance impedes venous return through the pulmonary vasculature resulting in upstream venous congestion (elevated CVP) and downstream restricted flow (decreased cardiac output), due to a “bottleneck” effect.<sup>2</sup>

Ureteropelvic junction obstruction (UPJO) is either an anatomical or functional anomaly, causing a reduction in the urine passage from the renal pelvis into the ureter. UPJO is usually congenital, resulting from the aperistaltic nature of a part of the ureter. Other causes are acquired, including crossing vessels, urolithiasis, strictures, polyps, adhesions and malignancy. It is usually diagnosed with prenatal ultrasound imaging. However, in rare cases the disease can remain clinically silent until adulthood. If left untreated, hydronephrosis, chronic infection, urolithiasis and progressive deterioration of renal function can result.<sup>3</sup> In this paper, we present the case of a 36-years-old man, with Fontan physiology and UPJO, treated laparoscopically in our Center. The changes

observed in the cardiovascular system of these patients are of such specificity and complexity that they make the performance of any laparoscopic procedure extremely challenging.

## 1.1. Case presentation

A 36-years-old man presented in our Center with right flank pain. He recalled recurrent urinary tract infections during childhood. Ultrasonography revealed mild hydronephrosis of his right kidney (the pelvis was distended but not the calyces). We decided to proceed with further workup for ureteropelvic junction obstruction, during which intravenous urography was consistent with a typical ureteropelvic junction obstruction pattern and a diuresis renal scan presented 39% uptake of the tracer and a GFR of 59 mL/min with delayed excretion (T1/2 22 minutes). Both blood (Urea and Cr) and urine tests were normal, while hypertension was excluded with successive measurements. As already mentioned, the patient has been diagnosed with single ventricle asplenia before birth and he has been subjected to the Fontan procedure when he was 3-years-old. As for all patients with ureteropelvic junction obstruction treated in our Center, our team decided to perform laparoscopic pyeloplasty, despite the demanding nature of this particular patient’s cardiovascular physiology. The operation was performed under low impact laparoscopy conditions, with the pneumoperitoneum pressure remaining stable at 8 mmHg throughout the procedure, so as to

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ensure that venous return to the heart is preserved at the highest level possible.

The basic steps of our technique were: 1) meticulous dissection of the right ureter up to the level of the ureteropelvic junction, 2) excision of the stenotic section of the right ureter, 3) spatulation of the ureter (Fig. 2), 4) placement of a self-retaining ureteric pigtail catheter, 5) anastomosis of the right renal pelvis with the spatulated right ureter (Fig. 3), 6) check of the anastomosis' watertightness with normal saline infusion into the bladder. In order to perform the operation, the contribution of the intensivists and of the interventional cardiologists of our hospital was necessary. The procedure was successfully completed in 95 minutes, while the blood loss was 45 ml. The postoperative course was uneventful. The patient remained in the hospital for 2 days and was discharged on postoperative day 3. The pigtail was removed 1 month postoperatively and no dilatation of the right pelvicalyceal system was observed on follow-up imaging. At 3 months postoperatively, a revision diuretic renal scan was performed, which was significantly improved. At 6 months' follow-up, the patient was completely symptoms-free.

**2. Discussion**

As already mentioned, Fontan physiology is characterised by increased intravenous pressure and decreased cardiac output. Insufflation of carbon dioxide into the abdomen with the resultant increase in the intra-abdominal pressure, the use of the reverse Trendelenburg position and the positive pressure ventilation, during laparoscopic procedures, can further compromise venous return. Moreover, hypercarbia raises the pulmonary vascular resistance, further reducing cardiac output.<sup>4</sup> In our case, the operation was performed under low impact laparoscopy conditions, with the aid of AirSeal® system, and thus the pneumoperitoneum pressure remained stable at 8 mmHg throughout the whole procedure, so as to ensure that venous return to the heart is preserved at the highest level possible. Furthermore, difficulty of bleeding control because of high CVP, congestive liver and liver cirrhosis (associated with Fontan physiology) can potentially make a laparoscopic approach extremely risky and challenging.<sup>5</sup>

Very few laparoscopic operations, in patients with Fontan physiology, have been described in the literature.<sup>4,5</sup> However, to the best of our knowledge, this is the first case of a urological laparoscopic

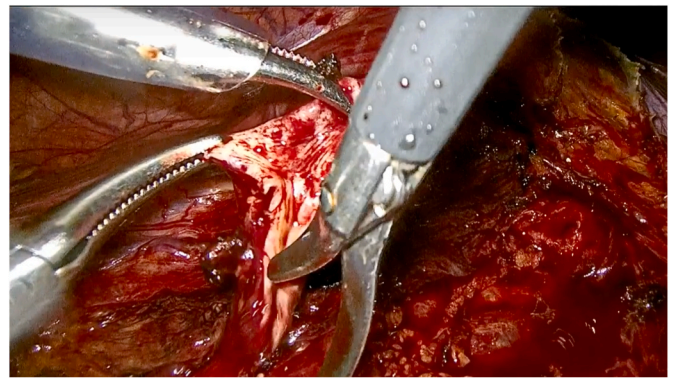


Fig. 2. Spatulation of the right ureter.

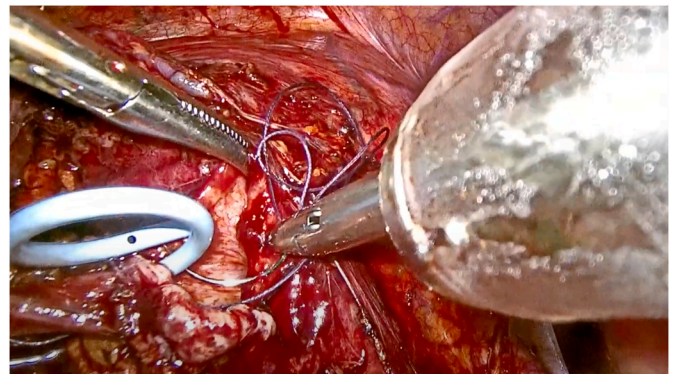


Fig. 3. Anastomosis of the right renal pelvis with the spatulated right ureter.

operation performed in a patient with Fontan physiology. Although any laparoscopic pyeloplasty represents a challenging reconstructive surgery, the elevated central venous pressure and the decreased cardiac output, which were associated with Fontan physiology, rendered this case very demanding and special. Another question is whether UPJO

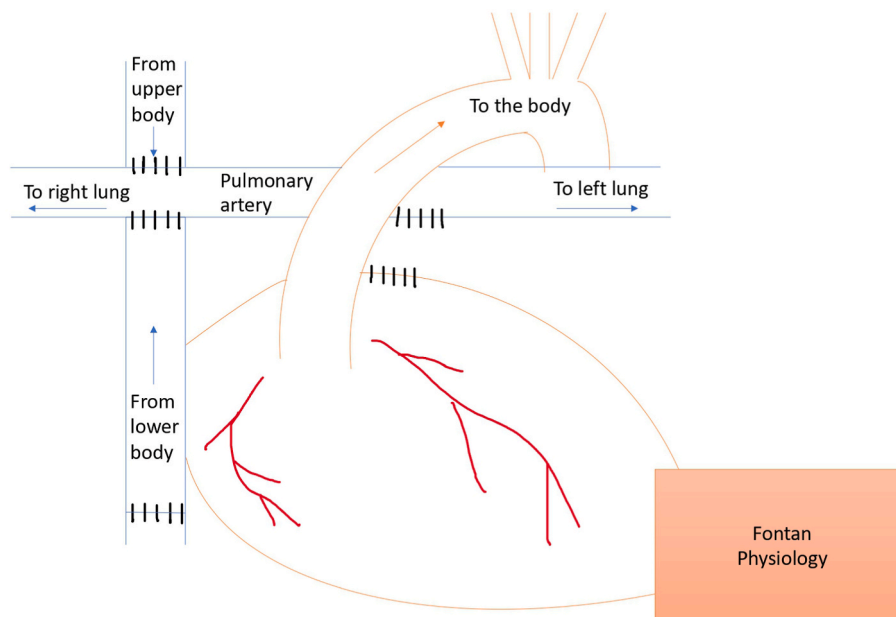


Fig. 1. Fontan Physiology. The systemic venous return is directly connected with the pulmonary arteries, resulting in passive filling of the pulmonary circulation without the need of a ventricular contraction. This modification prevents intracardiac mixture of arterial and venous blood, leading to increased oxygen saturation of arterial blood and protecting the single ventricle from volume overload.

could be associated with Fontan physiology. The patient with Fontan failure may have various problems, such as reduced exercise ability, ventricular dysfunction, dysrhythmias, cyanosis, ascites, hepatomegaly, cirrhosis, lymphatic system failure, venous thromboses and peripheral oedema.<sup>2</sup> Although extended search in the literature didn't end up with any positive results, we could not exclude it as a scenario. For example, increased central venous pressure and altered cardiovascular physiology could be related to a crossing vessel. However, in our case UPJO was idiopathic and thus a crossing vessel was not the cause of the stenosis.

### 3. Conclusion

Laparoscopic pyeloplasty for the treatment of ureteropelvic junction obstruction in patients with Fontan physiology, is an efficient and safe technique when performed in centers with extensive experience in laparoscopic procedures. The dynamic cooperation between different medical specialties (in our case between urologists, anaesthesiologists, intensivists and cardiologists) is critical for the success of such complicated procedures.

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### Declaration of competing interest

The authors declare no conflicts of interest.

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