

# Pyelolithiasis based on Ureteropelvic Junction Obstruction with Situs Inversus Totalis: Handling of a Child

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## Citation

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## Abstract

Dear Editor,

Four years old male patient who had been diagnosed as situs inversus totalis in another university hospital was admitted to our pediatric surgery clinics with haematuria complaints continuing for approximately 2 months. In physical examination, there was no abnormal finding. Plain radiography showed pyelolithiasis at the left-sided right kidney. Sonography, intravenous pyelography and retrograde pyelography were performed respectively to evaluate the stone etiology, and consequently ureteropelvic junction obstruction was diagnosed. Surgical treatment was planned for both obstruction and pyelolithiasis.

Before the operation, patient was premedicated with 0,5 mg/kg midazolam per oral. After adequate sedation, anesthesia induction within usual doses of propofol, fentanyl and vecuronium was applied. Patient was intubated with adequate tube. Sevoflourane and O<sub>2</sub>/N<sub>2</sub>O mixture was used for anesthesia maintenance. Pyelolithotomy with dismembered pyeloplasty was performed via left lateral flank incision. The only difficulty during the operation was the presence of the right liver lobe for the exposure and the operation was ended in 3 hours with minimum bleeding. We did not observe a problem during wake up and after the operation. Five days later, patient was discharged and his control examinations were totally normal.

All vertebrates have internal asymmetry along the left–right (L–R) axis (1). Embryologically, final asymmetry forms in 3 steps: First step is breaking of global embryonic bilateral symmetry and stabilizing this information in an organizing center. Second step is triggering the cascade of asymmetric gene expression and third step is recognition of the L–R information and translation into asymmetric organ morphology (1). In morphology, this mechanism comes out

in two types: 1- Asymmetry of unpaired organs such as the heart, liver, spleen, stomach, small bowel and colon, 2- Asymmetry of paired organs, specifically the lungs, the bronchi and the cardiac atrium appendages. A defect in global situs orientation, causing a complete mirror image is called situs inversus totalis (1). Each organ maintains their normal relative position with respect to another in situs inversus totalis (1).

Situs disorders are frequently associated with organ malformations and/or functional problems (1). The best known entity for situs inversus totalis with primary ciliary dyskinesia: Kartagener syndrome (1). Also some other diseases, particularly with kidney, were reported with situs inversus totalis. Renal cell carcinoma, renal dysplasia, cystic dysplastic kidneys, infantile nephronophthisis, renal hydatid cyst, Ivemark syndrome (involving kidney, pancreas and liver anomalies) and a new autosomal recessive syndrome (involving kidney, pancreas and lungs) were some renal diseases seen with situs inversus totalis (1,2,3,4). To our knowledge, our patient is the first one with pyelolithiasis and ureteropelvic junction obstruction. Therefore, we could not find a possible explanation of this coexistence in literature but assumed that the situation was only a sporadic association.

Therapy procedures, when needed, with situs inversus totalis patients are not well described. Rarity of the diseases associated with situs disorders may be the reason. There was only one report revealing intraoperative trouble with situs. Nayak et al reported that their patient with situs inversus totalis had an atypical cholinesterase which had been diagnosed during the anesthesia management (5). In their report they emphasized that situs inversus could be a warning signal for potential complications with the administration of succinylcholine (5). We suggested that

anesthesia of these cases was seemed to be a routine procedure but there were not enough patients in literature to propose a final decision for the simplicity of the management. On the other hand, there could be a positional difficulty during the operation because of the situs anomaly. In our experience, we realized surgical management of the patient might be shorter but in general, operation was not as difficult as our first impression. There were also no details for surgical procedures of these patients in literature.

In our letter, we aim to declare the first coexistence of situs inversus totalis and ureteropelvic junction obstruction with pyelolithiasis and share our experience of treatment. This report may facilitate the decisions for these types of patients in the future because neither anesthesia nor surgical managements are defined in details in the literature.

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