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Anaesthesia Section

# Anaesthetic Challenges and Management Strategies for Cystourethroscopy with Retrograde Pyelography and Left Ureteroscopic Surgery in a Patient with Kyphoscoliosis and a History of Operated Cystic Hygroma: A Case Report

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## **ABSTRACT**

Scoliosis is an idiopathic condition that predominantly affects females, with a ratio of 4:1, and an incidence ranging from 0.3-15%. Kyphoscoliosis, which involves both forward and lateral bending of the thoracolumbar spine, commonly results from idiopathic causes (70% of cases), with secondary causes including neuromuscular, congenital, or traumatic factors. The clinical significance of kyphoscoliosis lies in its potential to compromise respiratory mechanics and alter anatomical landmarks, posing challenges in perioperative and anaesthetic management. Addressing these challenges becomes more crucial when kyphoscoliosis is associated with other congenital anomalies, such as Cystic Hygroma (CH). CH is a common benign lymphatic cyst found in paediatric patients. Even after surgical intervention, CH can leave residual anatomical and physiological alterations that persist into adulthood, which may present anaesthetic challenges, including respiratory obstruction and the need to rule out additional congenital syndromes that necessitate comprehensive preoperative evaluation. This case report presents the anaesthetic challenges and perioperative considerations in a 18-year-old female with a history of operated CH and multiple congenital anomalies who is undergoing urological surgery. The presence of spinal deformity, restrictive lung disease, cardiac irregularities and prior surgical interventions necessitated a carefully tailored anaesthetic plan based on a multidisciplinary approach. The co-occurrence of congenital lymphatic and skeletal anomalies in this patient highlights the need for a multidisciplinary strategy, careful airway assessment and individualised anaesthetic planning to ensure optimal perioperative outcomes. This discussion explores the anaesthetic challenges and perioperative management in a case involving a child who underwent CH surgery, subsequently developed scoliosis in adulthood, and is now scheduled for cystourethroscopy with retrograde pyelography and left Ureteroscopic Surgery (URS).

Keywords: Lymphangioma, Pulmonary hypertension, Spinal deformities, Urologic surgical procedures

## **CASE REPORT**

An 18-year-old unmarried female patient presented with complaints of left flank pain, intermittent fever and chills, bedwetting on and off since birth and an inability to sense the urge to pass urine for the past two years. The patient also has bilateral lower limb dysplasia and was admitted for cystourethroscopy with retrograde pyelography and a left ureteroscopic procedure due to a left ureteric stricture. Her birth history includes a full-term normal vaginal delivery. She had a CH below the right scapula, for which she underwent corrective surgery twice, 17 years ago. She also had bilateral clubfoot that required corrective surgeries, ultimately leading to bilateral ankle disarticulation 14 years ago.

General and systemic examinations were normal, except for an irregular pulse of 74 beats per minute, an SpO<sub>2</sub> of 93% on room air, and reduced air entry at the lung bases bilaterally. The patient appeared apprehensive and had a low breath-holding time of less than 20 seconds. Airway assessment revealed a Mallampati Classification of II. On inspection, there was a hump on the right side of the scapula (15×12 cm) and another swelling immediately below that hump (12×10 cm) [Table/Fig-1].

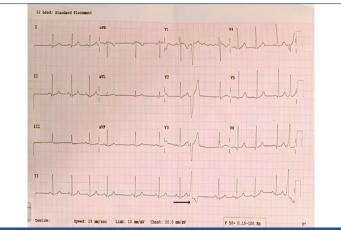
Laboratory parameters were normal, except for haemoglobin at 9.2 g/dL and white blood cells at 14,100/µL. The Montex test was negative. Chest X-ray showed increased bronchovascular markings, hilar opacities, dextroscoliosis of the lower dorsal spine with the apex



at the T11-12 level, and minimal dextroscoliosis of the lumbar spine with the apex at the L3 vertebra [Table/Fig-2]. Lumbar X-ray revealed minimal narrowing at the L4-L5 and L5-S1 levels. Spirometry indicated mild restrictive disease. The Electrocardiogram (ECG) revealed an irregular rhythm with occasional Ventricular Premature Complexes (VPCs) [Table/Fig-3]. Holter monitoring also showed VPCs. Two-dimensional echocardiography revealed an ejection fraction of 50%, trivial tricuspid regurgitation, moderate diastolic dysfunction and pulmonary hypertension. The patient received cardiology clearance with a moderate risk for non cardiac surgery.



[Table/Fig-2]: Chest X-Ray showing dextroscoliosis of the lower dorsal spine with the apex at the T11-12



[Table/Fig-3]: ECG showing an irregular rhythm with occasional Ventricular Premature Complexes (VPCs)

Ultrasound suggested bladder outlet obstruction and a possible left lower ureteric obstruction, which was later confirmed as stricture formation by CT-KUB. A consultation with pulmonary medicine, prompted by the low breath-holding time, resulted in clearance with a recommendation of moderate surgical risk, noting a mild restrictive pattern on Pulmonary Function Tests (PFTs). An orthopaedic consultation for scoliosis advised against any surgical interventions.

Authors conscientiously evaluated the patient to define the size and extent of swelling, assess the possibility of a difficult airway and evaluate the risks involved. Authors proceeded with the case under ASA 2E for cystourethroscopy with retrograde pyelography and the left Ureteroscopic (URS) procedure, with high-risk consent and ICU ventilator backup. A preoperative Arterial Blood Gas (ABG) analysis showed mild alkalosis with CO<sub>2</sub> retention and an SpO<sub>2</sub> of 93%. After administering antibiotic prophylaxis through a 20G intravenous catheter in the preoperative room, the patient was shifted to the Operating Room (OR). Routine monitors, including ECG, Non Invasive Blood Pressure (NIBP), and SpO<sub>2</sub>, were attached. As the patient appeared apprehensive, her spinal anaesthesia was counseled. However, while palpating the intervertebral space, she fainted, so 100% oxygen was provided with bag and mask

ventilation. Blood Sugar Level (BSL) was measured and found to be 84 mg/dL, and vital signs were noted, showing no bradycardia or hypotension.

As there was very little space for regional anaesthesia, general anaesthesia waspted. Since the patient could not walk and may have some component of myopathy, muscle flaccidity presented a risk for Malignant Hyperthermia (MH). Due to the gibbus below the right scapular area, we placed a wedge under the left side of her back to counter the asymmetry in her supine position. After premedication, anaesthesia was induced with Propofol (2 mg/kg), Fentanyl (2 mcg/kg), and Atracurium (0.5 mg/kg). Intubation was performed under video laryngoscopy using a cuffed PVC tube size 7. The placement of the tube was confirmed with capnography and the tube was subsequently fixed. In this case, succinylcholine and inhalational agents was avoided. Analgesia was administered in the form of intravenous Paracetamol (1 g). The patient was maintained on a mixture of oxygen and air in a ratio of 50:50, along with a propofol infusion (3-8 mg/kg) as a target-controlled infusion (TIVA), starting at a low dose. There was one episode of hypotension, which was managed with intravenous Mephentermine (6 mg). After spontaneous breathing attempts, the patient was reversed and extubated. The patient was then transferred to the Post Anaesthesia Care Unit (PACU) for monitoring and subsequently moved to the ward. She was discharged on the 5<sup>th</sup> day after drain removal, with a per urethral catheter in situ, which was removed on follow-up on day 14, and DJ stent removal was performed after six weeks.

### DISCUSSION

Kyphosis refers to a curvature of the spine that results in a rounded or hunched back. Scoliosis, on the other hand, is characterised by a lateral curvature of the spine exceeding 10°, often accompanied by a rotation of the vertebrae. In females, adolescent scoliosis is common. When there is kyphoscoliosis, securing the airway can be challenging, making it one of the most critical aspects of managing the case [1,2]. Understanding the anatomy of the spinal cord is vital, especially in the presence of kyphosis, scoliosis, or a combination of both. Particular attention must be given to the mobility of the cervical and thoracic spine, the presence of any abnormal anatomical structures and deformities such as a gibbus [3].

Kearney DJ and Davies SF, investigated pulmonary function in kyphoscoliotic patients without other cardiopulmonary diseases. Despite the lungs being functionally normal, kyphoscoliosis causes distortion of the thoracic cage, which decreases respiratory efficiency and increases the work of breathing. Severe, long-standing scoliosis is associated with significant ventilation-perfusion (V/Q) mismatch, leading to alveolar hypoventilation and  ${\rm CO}_2$  retention. This exacerbates the restrictive lung disease, contributing to chronic hypoxia and hypercapnia, and further increases the risk of pulmonary vasoconstriction, pulmonary hypertension and ultimately right ventricular hypertrophy and cor pulmonale. Therefore, it is imperative to carefully manage anaesthesia in such patients due to the risk of hypoxemia and respiratory complications [4,5].

The cardiovascular system often shows right ventricular hypertrophy and failure, secondary to pulmonary hypertension. Before finalising the anaesthesia plan, it is crucial to consider the multiple systemic issues associated with myopathy and kyphoscoliosis. These include the challenges of a difficult airway, restrictive pulmonary disease, and reduced cardiac output [6]. Spinal anaesthesia is almost always challenging in such cases, as palpation of anatomical landmarks becomes difficult and predicting the extent of the subarachnoid block is not easy. The severity of the disease can be assessed using Cobb's angle to classify pulmonary impairment as mild, moderate, or severe. These factors make general anaesthesia risky and present technical difficulties with regional anaesthesia due to the abnormal spinal curvature [6].

Congenital Hygroma (CH) is a congenital disorder that originates from the sequestration of lymphatic tissue from its sac [7,8]. Possible associated conditions with CH include haemodynamic alterations and cardiac dysfunction. A thorough preoperative evaluation is necessary to rule out associated congenital anomalies, define the extent of the swelling, assess the possibility of a difficult airway, and evaluate the risks associated with scoliosis. Spinal deformities present both functional and physical challenges for the patient and anaesthesiologists in planning the anaesthesia technique. Rosenberg H et al., reviewed the association between myopathies, metabolic disorders and the risk of MH [9].

The authors stressed the importance of a thorough preoperative assessment and individualised anaesthetic planning, especially for patients with musculoskeletal deformities like kyphoscoliosis, to minimise perioperative risk [9]. Despite being technically difficult, spinal anaesthesia is widely used due to the problems associated with the respiratory system [10]. In patients with multiple co-morbidities who are immobile, cannot walk and may have some component of myopathy, muscle flaccidity may pose risks of prolonged paralysis, hyperkalemia, rigidity, MH, cardiac arrest, rhabdomyolysis, or even death. When managing multiple co-morbidities, as in present case, the primary goals are to avoid myocardial depression, maintain normovolemia and optimal intravascular volume and ensure adequate systemic vascular resistance to achieve stable haemodynamics.

# CONCLUSION(S)

In patients with complex congenital anomalies like kyphoscoliosis and operated CH, anaesthetic management requires meticulous preoperative evaluation and individualised planning. The presence

of spinal deformities and potential airway challenges can limit the feasibility of regional anaesthesia and increase the risks associated with general anaesthesia. Multisystem involvement, such as restrictive lung disease and cardiac dysfunction, further complicates anaesthetic care. A multidisciplinary approach, vigilant intraoperative monitoring and preparedness for difficult airway and ventilatory challenges are crucial for a safe perioperative outcome. Tailoring the anaesthetic technique to patient-specific risks ensures better surgical outcomes and enhances patient safety.

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