

# Anticipated Challenges during Anaesthetic Management of a Patient with Kartagener Syndrome: Case Report

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## Abstract

The trait is Autosomal recessive. Primary ciliary dyskinesia is the major patho-physiology with Sinusitis, Bronchiectasis and Situs Inversus as the characteristic features. Preoperatively Respiratory and Cardiovascular system require special optimisation because of the presence of ciliary dysfunction, the management should be customised to avoid problems in the perioperative period. In present case few pre-emptive efforts like avoiding high FiO<sub>2</sub>, anticholinergics and thorough endobronchial suctioning utilizing fiberoptic bronchoscope before extubation helped us to avoid anticipated complications. Features important from anaesthetic view point include bronchiectasis and pan sinusitis causing airway obstruction, gas trapping and arterial hypoxemia that can later proceed to acute respiratory failure.

**Keywords:** Kartagener Syndrome; Situs Inversus; Bronchiectasis; Sinusitis.

## Introduction

The approximate incidence is 1:32000 individuals. The individuals present with male infertility and chronic and repetitive respiratory tract infections [1,2].

Respiratory and Cardiovascular examination requires special attention preoperatively. Patients may have productive cough, sinusitis, history of repetitive respiratory tract infections, otitis media, and infertility [3].

The main pathology is the underlying dysfunction of dynein arms in the cilia structure, damaging the coordinated movements of the cilia of the epithelial cells and mucociliary activity. Mucus retention occurs due to ciliary dysfunction leading to chronic respiratory infections and bronchiectasis ensues [4].

## Case Description

45 year old male patient, non-smoker, born to non-consanguineous parents, weighing 50 kg, carpenter by occupation presented to out-patient department with complaints of recurrent attacks of common cold, discharge and stuffiness of the nose. It was associated with cough with expectoration for past 10 years. He complained of exertional shortness of breath since last 3 years, and not having children despite being married for last 15 years.

Even during childhood, he used to develop cough, cold, rhinorrhoea, nasal blockade, and ear discharge frequently. He was prescribed anti-tubercular treatment, 5 years back but he had no relief. He was also under treatment of pulmonologist for recurrent LRTI since last one year. The patient was diagnosed to be a case of Kartagener

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syndrome and scheduled for functional endoscopic sinus surgery (FESS).

Vital parameters, Respiratory and Cardiovascular status was assessed Pre-operatively and found to be within normal limits. On physical examination Apex beat was felt in the right side in 5<sup>th</sup> intercostal space and Grade 2 digital clubbing was found. The.

On auscultation, bilateral basal wheeze and crackles were audible. The heart sounds were best heard on the right side of the chest. The patient had a peripheral oxygen saturation of 93%-94% on spontaneous ventilation with room air. The saturation improved to 100% on oxygen supplementation @ 2l/min, delivered through nasal prongs. Preoperative ABG was within normal limits (acceptable).

On evaluation, Electrocardiogram (ECG) showed evidence of dextrocardia. Chest X-ray postero-anterior (PA) view (Figure 1) showed cardiac apex and aortic arch on the right side, suggesting dextrocardia. The right sided stomach bubble was also noted.

CT Head and Neck was suggestive of sinusitis (Figure 2) with hypertrophic turbinate's and mucosal hypertrophy. Radiograph of sinuses showed mucosal thickening in maxillary sinuses.



Fig. 1:

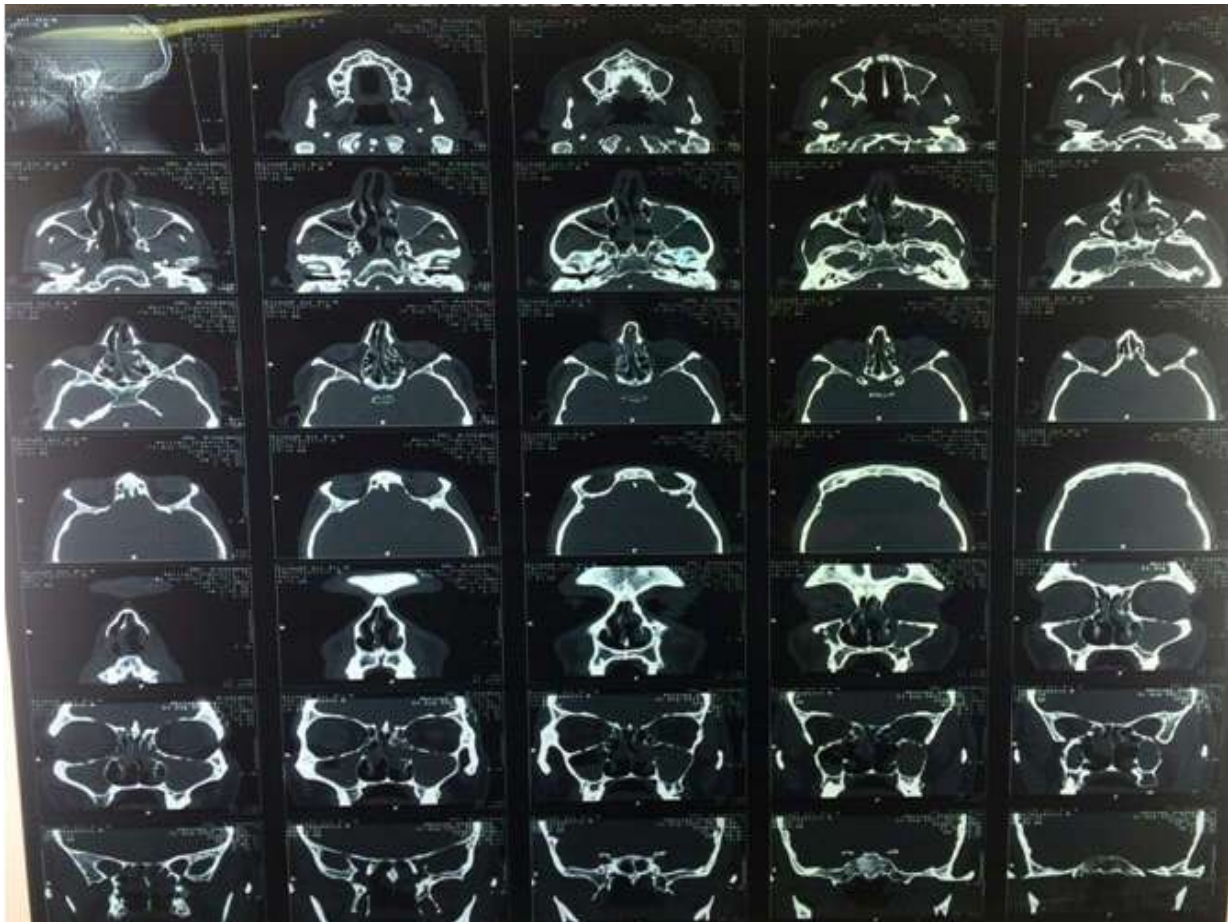


Fig. 2:



Fig. 3:

An ultrasound abdomen was done but it did not reveal any abnormality. Contrast-enhanced computed tomography (CECT) chest (Figure 3) also revealed dextrocardia, few nodular opacities were seen in right lower lobe, which were suggestive of bronchiectasis.

The semen analysis revealed azoospermia.

Patient had undergone FESS in past under local anaesthesia but this time he requested GA.

Pre-operatively chest was optimized with antibiotics, bronchodilators, steam inhalation, chest physiotherapy and nebulisation. Informed written consent was obtained.

Patient was told to be kept nil per oral after 12 midnight. Medications were continued till the day of surgery. Multimodal monitors were attached. 18 G cannula was inserted in left forearm for iv access.

Patient was premedicated with inj. Ondansetron 0.15mg/kg iv. and Inj.Fentanyl 1 microg/kg/iv and preoxygenated with 100% oxygen via face mask. Patient was induced with Inj. Propofol (2mg/kg) and vecuronium bromide 0.8mg/kg and intubated with 8.5 CETT. Anaesthesia was maintained with 1% isoflurane in 40% O<sub>2</sub>-N<sub>2</sub>O mixture, isoflurane and intermittent doses of muscle relaxant. Throat packing was done.

Before extubation thorough endobronchial suctioning was done using FOB which was inserted through endotracheal tube. Still the auscultation revealed wheezing and hence bronchodilator therapy was administered. Patient was extubated and shifted to ward.

Inj.Paracetamol 1 gm with Inj.Tramadol 50 mg was given 8 hrly were advised for Post-operative analgesia. The postoperative period was uneventful

although the patient required frequent endotracheal aspirations and his bronchodilator therapy was continued.

## Discussion

Few steps are crucial in preoperative optimization like Respiratory physiotherapy, postural drainage, antibiotic therapy, and bronchodilators. Cautions to be exercised during anaesthesia are as follows- Firstly, the ECG electrodes and defibrillator paddles are to be placed as a mirror image of their normal positions.

Secondly, there are increased chances of advancing of the ETT to the left lung because of Situs Inversus.

In case of inversion of great vessels, left internal jugular vein should be preferred for cannulation, as it will give direct access to right atrium and will also avoid injury to thoracic duct [5].

Ciliary dysfunction increases the probability of perioperative pulmonary risk increases and if feasible, regional anaesthesia should be preferred over general anaesthesia [5] but that was not possible in our case.

In premedication theoretically cough suppressants or drugs that depress ventilation or ciliary activity should be avoided [6]. All these were taken care in our patient by giving bronchodilators, steroids, and short acting opioid, avoiding anticholinergics in premedication.

Choice of fentanyl as narcotic, provided basal analgesia for the period of surgery without the undesirable ventilatory side effects in the postoperative period.

Inhaled medical gases, high O<sub>2</sub> concentrations, and anaesthetic agents can affect the already decreased mucociliary functions, thus we deliberately avoided high FiO<sub>2</sub> in this case.

Preoperative chest physiotherapy and antibiotic therapy is also to be considered [7]. Both were taken care preoperatively.

Viscous mucus plugs may cause respiratory distress post-operatively therefore bronchoscopic aspiration can be beneficial before extubation.

Functional Endoscopic Sinus Surgery (FESS) and the formation of a nasal antral window below the inferior turbinate may afford a transient improvement in upper and lower respiratory tract symptoms.

There was significant improvement seen on follow up after endoscopic sinus surgery and the incidence of recurrent infections also reduced drastically. Due to abnormal neutrophil chemotaxis in such patients it is very important to maintain Antisepsis during any interventions like epidural, central venous catheterization and aspiration of the endobronchial tube [8].

Although all possible precautions are taken, infections, respiratory infections can occur and increase morbidity and hospital stay.

## Conclusion

The most challenging aspect in the anaesthetic management of Kartagener syndrome is avoiding pulmonary complications resulting from cilia dysfunction and bronchiectasis. It is prudent to remove the mucoid secretions with effective aspirations. It is important to remember that drugs causing respiratory depression should be avoided and main emphasis should be on maintaining Oxygenation. Using regional blocks for pain treatment if possible, and securing postoperative respiratory support.

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