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Anaesthetic management of a patient with Pectus Carinatum for ophthalmic surgery

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Abstract
Pectus carinatum is a chest wall deformity that occurs less frequently. Anaesthesia in such patients may be complicated by associated systemic disorders such as Marfan's disease, homocystinuria, Prune belly syndrome, Morquio syndrome, Osteogenesis imperfecta, Noonans syndrome, mitral valve prolapse, scoliosis or other collagen vascular diseases. Though pectus carinatum is widely reported, the literature on anaesthetic management is sparse.

Keywords: Pectus carinatum, restrictive lung disease, Anaesthesia, Vitreo retinal surgery

Introduction
Pectus carinatum (PC), a genetic disorder, is a chest wall deformity characterized by anterior protrusion of the sternum due to overgrowth of costal cartilages and predominantly affects males [1, 2]. This report describes the successful management of a patient with Marfans syndrome, dorsal scoliosis and pectus carinatum for an ophthalmic surgery.

Course in the hospital
A 23 year old male patient weighing 47 kg presented to our institution with retinal detachment and was scheduled for vitreo retinal surgery under general anaesthesia. Patient was a known case of Marfan's syndrome and had limitation of physical activity due to pectus deformity.
On examination the patient was moderately built and had obvious wasting of muscles. The pectus carinatum deformity was quite marked (Fig.1, 2). The cardiovascular and respiratory system examination was unremarkable.

The haemogram and blood chemistry was normal
CXR showed pectus carinatum with mild dorsal scoliosis.
X-Ray of lumbar spine revealed lumbar scoliosis with concavity to the left.
Pulmonary function tests with spirometry revealed mild restrictive lung disease. Echocardiogram was normal.

Anaesthetic management
The patient was fasted as per universal fasting guidelines. Chest physicians and cardiologist's opinion was obtained. Patient was premedicated with I.M. Glycopyrrolate 0.004mgs/kg one hour preoperatively. Standered monitoring was applied and IV access was established with 20G cannula. 0.9% DNS was connected.
Anaesthesia was induced with I.V. Pentothal 5mg/kg and ventilation was assisted with O2, N2O and Sevoflurane. After adequate depth was achieved, trachea was intubated with 7.5mm ID cuffed endotracheal tube. Correct placement of the tube was confirmed by auscultation and EtCO2 trace and value. Analgesia was maintained with I.V. Fentanyl 1.5 mcg/kg and a subtenon's block with 0.5% Ropivacaine.

As patient had obvious muscle wasting, neuro muscular blocking agents were avoided and anaesthesia was maintained with volatiles agents, I.V analgesics and regional anaesthesia. Intraoperatively vitals remained stable for the duration of surgery which lasted for about two and a half hours (Fig.3). At the end of the surgery patient was extubated after complete return of reflexes and completely awake. He was observed in postoperative ward for two hours and shited to the ward.

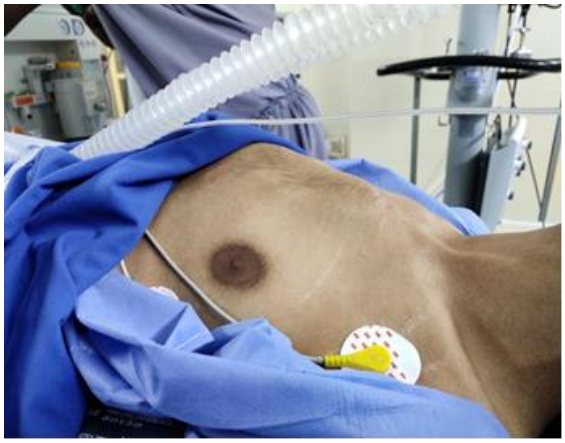

Fig 1: Pectus Carinatum

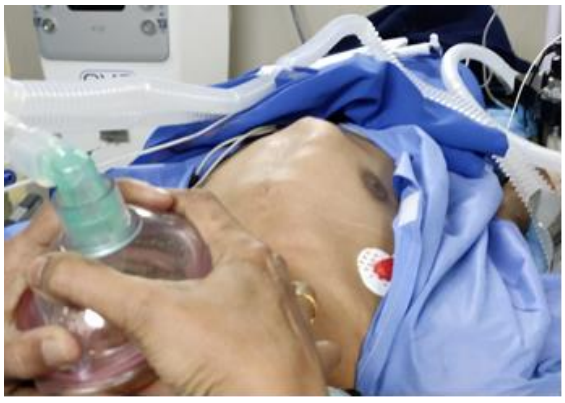

Fig 2: Pectus carinatum

Fig 3: Intra operative vitals

Discussion

Pectus carinatum or pigeon breast is a congenital protrusion deformity of the chest wall. This name is derived from a Latin word that means chest with a keel. It is an uncommon entity [3, 4]. It is six times less frequent than pectus excavatum [5]. The exact aetiology is unknown. However, there are many theories propounded. Overgrowth of costal cartilages with forward protrusion of manubrium, gladiolus and xiphoid is the most accepted one. There may be an association with other conditions like Marfan's syndrome, Poland syndrome or scoliosis. Pectus carinatum is estimated to occur in up to 0.06% of all live births, with an incidence of approximately 1 per 1000, seen in teenagers.

The deformity may present at birth but is usually evident in mid-childhood. However, it becomes most prominent during the adolescent growth spurt and most children are brought between eleven to fifteen years of age. Males are four times more affected than females [5]. There are three types of the deformity, namely, chondromanubrial, chondrogladiolar and a lateral variety which is rare [6]. In the chondrogladiolar PC, the chest cage is flexible and highly complaint, but in chondromanubrial PC, the chest is rigid and non-complaint. Unilateral outgrowth of the costal cartilages due to sternal rotation causes asymmetrical PC that is a variant of chondrogladiolar type. In the lateral type, it is the protrusion of costal cartilages which dominates the clinical picture.

The symptoms are owing to a rigid chest wall with an anteroposterior diameter which is almost fixed in full inspiration, arterial hypoxaemia and consequent cor pulmonale. As the lungs lose compliance, the incidence of emphysema and frequency of respiratory infections increases. Patients may present with retarded growth, exertional dyspnoea, asthma, palpitations and chronic dyspepsia. It has been suggested that PC leads to increased residual volume, decreased vital capacity and reduction in chest movements during respiration. Pulmonary function restriction is partly due to pectus and partly due to kypho scoliosis.

Thus, a thorough pre-anaesthetic evaluation, is mandatory for successful management of this rare condition with restrictive lung disease and reduced lung compliance. If such patients are found to have muscle weakness, it is preferable to avoid neuro muscular blocking agents especially in a single specialty set up. A combination of general and regional anaesthesia helpful even in long duration surgeries.

Financial Interest: None
Conflict of Interest: None

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