



## PERIOPERATIVE ANAESTHESIA CARE : IN POLAND SYNDROME - A RARE CASE REPORT.

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### KEYWORDS :

#### INTRODUCTION

Poland syndrome (PS) is a rare musculoskeletal congenital anomaly with variable presentations . Incidence is 1/30,000 live births, more often seen in males than females (3:1). Several theories have been proposed regarding the genesis of PS as vascular disruption theory “ The subclavian artery disruption sequence”. Clinical features include cardiovascular (dextrocardia, atrial septal defects) musculoskeletal (club foot, toe syndactyly, hemi vertebrae and scoliosis) genitourinary (renal aplasia and hypospadiasis) hemopoietic (spherocytosis and myelogenous leukemia, lymphoma) abnormalities. Absence of sternal head of pectoralis major muscle is an essential feature of PS and most often it manifests as ipsilateral thoracic cage and upper limb anomalies. Clinical features are phenotypically variable and manifestations are often subtle in infancy and childhood delaying their diagnosis and plan for intervention. PS patients does not always have functional defects but they present for cosmetic surgical repair where the safe anaesthetic intervention need to be planned .

#### CASE

A 20y old female patient (height 145cm, wt(42kgs) with left sided Poland's syndrome presented with complains of absence of left breast development scheduled for fat transfer breast reconstructive cosmetic surgery , In pre anesthetic check up patient found to have facial asymmetry present ( left side buccal fat absent ) left sternocleidomastoid muscle atrophy present with facial tilt towards left side with limited flexion of neck noted. Paradoxical respiratory movement not seen on auscultation respiratory system and cardiovascular system found no abnormalities . In cxr no rib cage abnormality present all routine investigations (cbc), RFT with serum electrolyte are normal. Also echocardiography of patient done –normal. Family history is not significant, patient have no previous history of surgery. Patient was informed about the type of anaesthesia and the complications associated it. Consent was taken . Patient was kept on fasting for 6hr before surgery .

On the day of surgery patient was secured with 18g IV cannula and premedicated with midazolam and taken on operating table. In operation theatre A difficult airway cart with different size airways ,laryngoscopic blade ,endotracheal tube , along with video laryngoscopy. All vital monitors attached (ECG, oxygen saturation ,NIBP).Preoxygenation started with 100%oxygen for three minutes . Anaesthesia induced with titrated dose of injection .Fentanyl 2mcg/kg and injection Thiopentone 5mg/kg , Bag and Mask ventilation done no difficulty present ,injection atracurium 0.5mg/kg was given and under direct laryngoscopy airway secured with ETT 6.5mm ID cuffed tube bilateral air entry checked equal and adequate .EtcO<sub>2</sub> graph appeared and ETT fixed. Intra operatively for maintenance of anaesthesia nitrous oxide 50% was give along with adequate hydration and analgesia maintained. Surgery was done in 2 hours of time fat transfer was done from bilateral thigh and abdominal regions. Patient was extubated awake after adequate return of neuro muscular power with injection neostigmine 0.05mg/kg and glycopyrrolate 0.005mg/kg. patient was stable hemodynamically through out the surgery without any adverse events of desaturation . Post operatively patient was

maintained of pain free and comfortable and discharged with uneventful hospital stay.



Fig 1. Strenocleidomastiod Atrophy

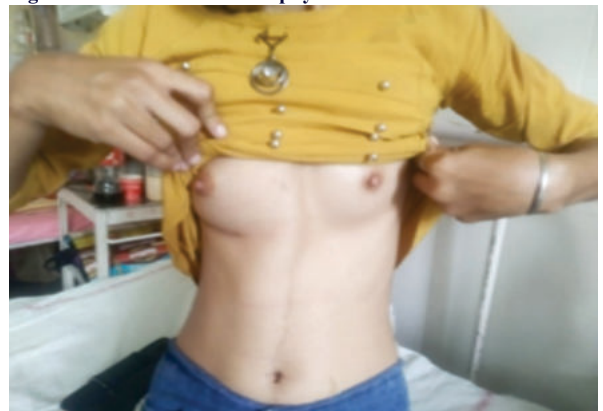


Fig.2 Breast Deformity With Facial Tilt.

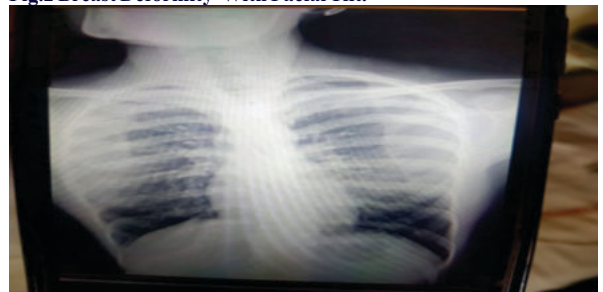


Fig 3. Cxr With Abnormal Straight Curvature Of Clavicle.

## DISCUSSION

Patients with Poland syndrome are associated with musculoskeletal and congenital anomalies that have multiple implications in securing airway and safe anaesthetic management of patient. In 1841, Alfred Poland reported first case of Poland syndrome absence of pectoral muscle alone was described earlier. Syndactyly was described to be associated with absent pectoral muscle by Patrick Clarkson and he coined the term "Poland syndactyly". Later it was Thompson who identified and summarized complete syndrome and features. To describe Poland syndrome several theories and etiological factors have been suggested as congenital disorders and birth defects occur due to an error in morphogenesis resulting in malformations and deformations. Embryonic vascular disruption sequence and the incidence was reported to be 0.1/1000 infants born with more than one structural anomaly. Other hypothesis include maternal smoking and cocaine abuse.

Pre operatively patient should be assessed system wise to rule out the associated anomalies and comorbidities present for further management. All baseline investigations to be done, CXR, ECG, 2DECHO to be done to rule out cardiopulmonary anomalies. A detailed airway examination should be done to anticipate difficult airway management. The most common surgeries for Poland syndrome that require anaesthesia were cosmetic surgeries such as breast reconstruction and usually have no vital functional impairment and surgical interventions are usually performed in late adolescence. Surgical reconstruction during early adolescence may have extremely positive impact on correct stabilization of body image and quality of life in young patients, In this surgery General Anaesthesia is first choice. The patients with Poland syndrome and some other congenital diseases show risk of developing malignant hyperthermia due to musculoskeletal abnormalities, so in this patients succinylcholine and maintenance on high inhalational anaesthesia is contraindicated. In the present case injection atracurium is used for muscle relaxation and low dose of inhalational anaesthetic agents were used, along with vital monitoring ( ECG, SpO<sub>2</sub>, HR) continuous temperature and Etco<sub>2</sub> monitoring was done to prevent the risk of malignant hyperthermia.

Patients with Poland syndrome can be asymptomatic and have paradoxical breathing, but in present case paradoxical breathing is absent. Intra operatively controlled positive pressure ventilation was given all over the procedure as uncontrolled spontaneous ventilation during general anaesthesia may cause cardiopulmonary collapse. Post operative pain management is important and can be given as regional anaesthesia are systemic with iv drugs (opioids and nsaid) risk benefits should be explained to patient before giving regional anaesthesia and higher dose of systemic analgesia.

## CONCLUSION

A cautious Anaesthetic management with proper pre operative assessment, adequate preparation and planning for management of difficult airway should be done. Intra operatively general anaesthesia with controlled ventilation is safely used for patients with adequate analgesia, hydration and continuous temperature management should be done to prevent patient risk of malignant hyperthermia. Post operative pain management should be adequate until proper recovery of patient.

## REFERENCES

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