






Case Report

Poland syndrome: A case of difficult weaning of ventilation after surgery

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ABSTRACT

Introduction: Poland syndrome is a rare congenital anomaly that occurs in 1 in 30,000 children. It is characterised by unilateral underdevelopment or lack of the chest wall muscle that is apparent at birth. Rarely is it discovered to involve a bilateral chest muscular deficit. Poland syndrome presents in a wide variety of ways, which can often make diagnosis challenging.

Aim: We would like to highlight the rarity of Poland syndrome, and the challenges of post-operative recovery, primarily the ventilation.

Case study: We presented a case of a 44-year-old female with non-investigated chest wall deformity, whom required emergency surgical intervention under general anaesthesia. Post-operatively, she required prolonged ventilatory support, and was then diagnosed with Poland syndrome.

Results and discussion: The patient had undergone an emergency laparotomy and left lateral hepatectomy for left hepatolithiasis with liver abscess. Post-operation, she required prolonged ventilation, with gradual weaning of oxygen. Imaging studies showed absence of left pectoralis major, thus a diagnosis of Poland syndrome. Gradually she was weaned off and had a successful post operative recovery.

Conclusions: Poland syndrome is a rare congenital abnormality. In patients undergoing general anaesthesia, the normal mechanics of breathing while ventilated are absent. Their recovery is longer and requires prolonged ICU admission.

1. INTRODUCTION

Poland syndrome is a rare congenital anomaly that occurs in 1 in 30,000 babies.¹ It is more common in males as compared to females. It is a combination of multiple chest wall anomalies which occurs with the pectoralis major, and it may include other defects such as hypoplasia of breast, absence of ribs, and in some cases syndactyly. The common feature is the occurrence of these anomalies on the ipsilateral side of the deformity. Despite the absence of chest wall muscle, majority of patients with Poland syndrome have no respiratory difficulties.¹ Majority of patients present with aesthetic complaints, and are the main reasons requiring surgical correction.

The etiology of developing Poland syndrome is unclear, most cases are sporadic. However, there are three other hypotheses about the origins of Poland syndrome, including genetic, teratogenic, or vascular impairment during foetal development.³

2. AIM

In this paper we present a case of a female patient who underwent an emergency operation under general anaesthesia and required prolonged ventilation and weaning post-operatively. We would like to highlight the dilemma of managing patient with Poland syndrome in view the chest wall deformity that made the post-operative recovery more challenging.

3. CASE STUDY

A 44-year-old female was admitted to our surgical unit for left hepatolithiasis with liver abscess. She had presented with right hypochondriac pain and fever for 3 days. There was no history of jaundice during the current admission. She had past medical history of choledocholithiasis requiring endoscopic retrograde cholangiopancreatography (ERCP).

On examination, she was febrile and septic looking. Per abdomen examination revealed tender hepatomegaly. She had absence of left chest wall, which was previously diagnosed as simple breast asymmetry (Figure 1) and no further investigations had been carried out. CT scan showed hypoplastic left 3rd and 4th rib, left breast and absence of pectoralis muscles, corresponding with Poland syndrome (Figure 2).

Due to worsening of her clinical condition despite antibiotic treatment, the managing surgeon decided to perform emergency laparotomy and left lateral hemihepatectomy.

Post-operatively, she was admitted to ICU after failure of extubation as she was unable to maintain good saturation. Throughout her ICU admission, there was difficulty weaning off her ventilation, with multiple changes to her ventilator settings. With gradual ventilatory weaning, patient made a successful post-operative recovery. She was successfully extubated at day 7 post-operatively and transferred out from ICU at day 9. Subsequently, patient was discharged well at day 13.



Figure 1. Asymmetrical breast and chest wall.



Figure 2. Absence of left pectoralis major muscle in CT scan.

4. RESULTS AND DISCUSSION

Poland syndrome is a rare congenital anomaly that occurs in 1 in 30,000 babies. It is a combination of multiple chest wall anomalies which occurs with the pectoralis muscle, and it may include other defects such as hypoplasia of breast, absence of ribs, and in some cases syndactyly. Majority of cases are ipsilateral, with very few being reported to occur bilaterally.¹

Typical presentation of Poland syndrome involves complaints regarding the aesthetic appearance of the chest, however they will rarely present with respiratory difficulties. Our patient had noticed the breast asymmetry since childhood, but since it didn't disturb her daily life work (and due to religious issue) she refused to get further intervention for the condition.

For clinical diagnosis, there is asymmetry of the chest wall, the specific characteristic will be absence of the sternocostal head of the pectoralis major muscle. For radiological diagnosis, USG is the first line modality to be used as it can detect absence of soft tissue and chest wall deformities. In the cases of severe Poland syndrome, CT scan or MRI is done for a more precise depiction of the chest wall deformity, bone anomaly and vasculature.²

In Malaysia, Poland syndrome is rare, with no cases being reported therefore the incidence is underreported and underdiagnosed. Most patients are asymptomatic, therefore other differential diagnosis was given and no further investigations were carried out.

Regarding this case, the patient had been misdiagnosed before the current admission and no further investigations were done regarding the asymmetry of the chest. Only the current admission CT scan revealed the absence of chest wall muscles thus the diagnosis was pointed towards Poland syndrome. However, after she had undergone a procedure of a prolonged time under general anaesthesia, difficulty arose in post-operative ventilation. Patients on ventilatory support without chest wall deformities can synchronise their respiratory muscles with ventilator settings, and as weaning occurs they are able to regulate their breathing.⁴ In Poland syndrome, due to the absence of pectoralis muscles and rib deformities, there is poor development of subatmospheric pressure in the thorax. In combination with paradoxical respiration, it causes inadequate ventilation and will lead to hypoxia.⁵

Other than the absence of muscle, there is also the presence of rib agenesis, such as in this patient. Due to the respiratory difficulties and prolonged ventilation, there may be role for rib reconstruction to improve the chest movement and her post-operative outcome, however since she had recovered, this operative management was not taken into consideration during this admission.⁶

Evaluating the case retrospectively, there are multiple ways to assess defects in the respiratory tract. The simplest form of respiratory assessment which is available in hospitals include the spirometry. Studies have shown that early assessment with a spirometry can help detect with early respiratory irregularities.⁷

In regards to the choice of anaesthesia, if the diagnosis of Poland syndrome had been made, adequate preparation can be made to reduce the risk of post operative recovery. Ideally regional anaesthesia is the best modality, but in this case patient requires general anaesthesia. As reported by Kumar et al., the choice of total intravenous anaesthesia and non depolarising muscle relaxants, and other modalities to reduce the risk of developing malignant hyperthermia.⁸

5. CONCLUSIONS

Poland syndrome is rare, and patients are asymptomatic. However if they undergo surgery with prolonged general anaesthesia, their recovery will hindered due to the loss of normal breathing mechanism, thus all patients with chest wall deformity should undergo further investigation, and subsequent treatment if warranted.

Conflict of interest

No conflict of interest.

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