

Poland syndrome: a case report

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DESCRIPTION

Poland syndrome (PS) is a rare chest wall developmental anomaly characterised by ipsilateral agenesis/hypoplasia of the sternocostal head of pectoralis major, hypoplasia of nipple or breast, absence of subcutaneous fat, multiple rib abnormalities, elevated and rotated scapula (Sprengel deformity) and ipsilateral digit abnormalities (brachydactyly, syndactyly). These findings vary and all are rarely found in a single individual.

Our case involves an 8-year-old boy with no medical history who presented for evaluation of chronic rhinitis. There was no breathing or cardiac complaints on review of systems. Physical examination showed chest asymmetry with right anterior chest wall depression and flattening of the right pectoral region with displaced nipple (figure 1). Abduction of the shoulders showed absence of the sternocostal head of pectoralis major. Hand examination did not show any signs of ipsilateral digital abnormality.

PS was first described by Alfred Poland in 1840. Aetiology of PS remains unknown. It is rarely genetically inherited and is regarded as a sporadic event. It has been suggested that during the sixth week of gestation, the injury occurs due to regional vascular defect of the subclavian artery.¹ It is a period associated with splitting of the two heads of pectoralis major and the development of tissues between the digits.

Treatment is primarily reconstructive surgery depending on the severity of the malformation, gender and patient preference. Generally, the reconstructive path is complex and must be performed stepwise because the malformation



Figure 1 Chest asymmetry with right anterior chest wall depression and flattening of the right pectoral region with displaced nipple.

Patient's perspective

My son was first diagnosed with Poland syndrome as an infant. Naturally, being his mother, I searched in medical journals and books and anything published on the computer. I must say that I was truly disappointed. There isn't a lot of information about and I'm assuming it's because research was limited. I would also like to point out that I could not find one photograph of what Poland syndrome looked like. Your interest in this will help so many that don't know what I know, and it's very limited. Good Luck to you, Dr Gonzalez. Please remember to send me a copy of your article.

Learning points

- ▶ Poland syndrome is a developmental disorder characterised by hypoplasia of the sternocostal head of pectoralis major, ipsilateral digit abnormalities (such as brachydactyly, syndactyly), nipple/breast and other chest wall and scapula abnormalities.
- ▶ Aetiology is unclear but is thought to be secondary to vascular defect of the subclavian artery during the sixth week of gestation.
- ▶ Surgical planning for patients with Poland syndrome is characterised by a personalised approach, which must consider patient's physical and mental aspects.

involves alteration of different tissues (skin, subcutaneous tissue, muscle, breast). Cardiac or respiratory complaints should alert the healthcare provider for further work-up. Left-sided PS has been associated with dextrocardia particularly in patients with agenesis of two or more ribs.² The 'vascular theory' has been proposed for other variants of this syndrome such as Mobius and Klippel-Fiel.¹ Several cases have described an association with neoplasms such as neuroblastoma, leukaemia, leiomyosarcoma, breast cancer and Wilms tumour.³

Reconstructive surgery is planned for this patient after growth completion. The patient has remained asymptomatic and will follow up with cardiothoracic surgery after growth completion.

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