

## IMAGES IN CLINICAL PRACTICE

### UNILATERAL FLAT CHEST WALL IN A CHILD

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An 8-year-old boy, second born to non-consanguineous parents presented to our hospital with complaints of a unilateral flat chest wall (Figure 1). There was no significant family history and the antenatal history was uneventful. On examination, his chest wall was asymmetric with right hypoplasia. The right axillary fold was absent. The right shoulder was placed a little lower and the right areola and nipple were placed a little higher than the left side. His shoulder adduction was slightly limited with a power of 4/5. On palpation, the pectoralis major muscle was absent on the right side. The pectoralis minor and serratus anterior muscles were present. The upper and lower limbs were normal on both sides. He did not have any skeletal abnormality. His cardiovascular and respiratory system examinations were normal. Radiological evaluation of the chest and echocardiography of the heart were normal.

**Figure 1.** Flat chest wall on Right side.



#### What is the diagnosis?

Poland syndrome (PS) was first described by Alfred Poland in 1840.<sup>1</sup> PS is characterized by the unilateral absence of the pectoralis major muscle. Hypoplasia or aplasia of other muscles like serratus anterior, latissimus dorsi and external oblique have also been described. PS is associated with other skeletal and visceral anomalies.<sup>2,3</sup> PS is usually unilateral, occurs

more in males and is more commonly seen on the right side. Though the etiology of PS is unknown, it has been postulated to be due to an interruption of blood supply in the subclavian artery during the embryonic stage of development.<sup>4</sup> The interruption in one or more branches of the subclavian artery leads to the associated anomalies. Bavinck and Weaver introduced the term "Subclavian Artery Supply Disruption Sequence" and common pathogenesis for Poland syndrome, Moebius syndrome and Klippel-Feil syndrome was proposed.<sup>4</sup> The involvement of the breast in the syndrome ranges from mild hypomastia to amastia. The rib anomalies that are commonly observed were the absence of costal cartilages in 2, 3 or 4th ribs which can lead to herniation of the lung tissue. The skeletal abnormalities that were commonly reported include elevation of the shoulder blade (Sprengel deformity), shortening of the arm and/or forearm, brachydactyly and syndactyly.<sup>2</sup> Cardiac anomalies like dextrocardia are commonly associated when the left side was involved. Many malignancies were reported in patients with PS including leukemia, lymphoma, lung carcinoma, gastric carcinoma and head and neck tumors.<sup>5</sup> In the present case, the absence of the pectoralis major muscle alone without any other abnormalities was observed. No surgical treatment was offered and the family was counseled. The management of PS depends on the extent of anomalies and functional limitations. The Italian Association for Poland Syndrome has provided best practice recommendations for the management of patients with PS.<sup>6</sup> The treatment of patients with PS is mainly done for aesthetic reasons. For chest deformity, silicon prosthesis is safely used.<sup>7</sup> In females, the reconstruction of the breast will be advantageous. Corrections of other deformities, like syndactyly, are also indicated whenever possible. Also, children with PS should be given psychological support that will improve long-term outcomes.

#### Compliance with Ethical Standards

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**Conflict of Interest:** None

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