

Cleft Hand with Ipsilateral Chest Deformity – Poland's Syndrome

R.K. Gang¹ and S. Makhlof

¹ Department of Plastic and Hand Surgery and

² Department of Orthopaedics, Faculty of Medicine, Arab Medical University, Benghazi/Libya

Summary. A case of Poland's syndrome in an adult is described with an additional feature of absence of flexor tendons of the little finger.

Key words: Poland's syndrome.

In 1841 Alfred Poland described the association between simple syndactyly and ipsilateral absence of pectoralis major and minor muscle in a cadaveric dissection. Since that time several reports have appeared in the literature [1–5] describing the aplasia of pectoral muscle, with a varying degree of congenital abnormalities on the same side; this has been termed Poland's syndrome. The main characteristic features of the syndrome are unilateral simple syndactyly, usually affecting all four fingers, a hypoplastic hand, and ipsilateral absence of sternocostal portion of the pectoralis major with or without associated chest wall and mammary gland defects.

It is surprising, however, that there are only two reports [7, 8] describing the abnormality of the tendons in addition to hypoplastic hand and forearm. Few patients are reported [1, 2, 6, 9, 10, 13] to have more severe limb deformity than those classically described. We describe an additional feature, in a classical case of Poland's Syndrome, of absence of flexor tendons of the little finger.

Case Report

A male patient with deformity of the right hand was first seen at the age of eighteen years for the purpose of obtaining a medical report to obtain a driving licence. On examination the right upper extremity was found to be hypoplastic and the right arm and forearm were deficient in length by 1.2 and 1 cm compared to the left side. Elbow and shoulder movements were normal. Only the thumb and little finger were present. The middle three fingers were absent (Fig. 1). There was no active movement at the interphalangeal and metacarpophalangeal joints of the little finger but passive movement was possible. The interphalangeal joint of

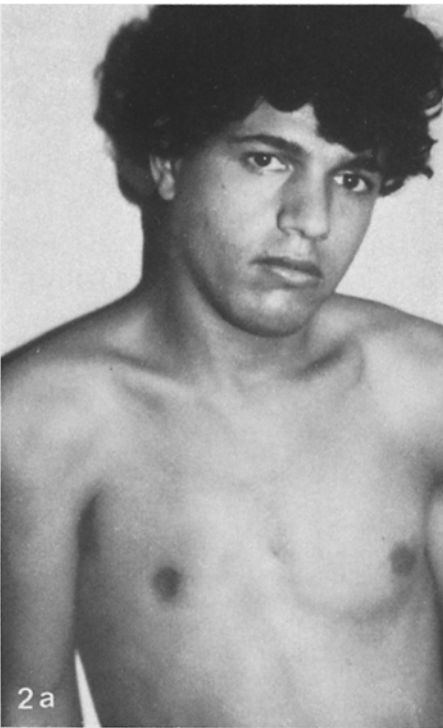


Fig. 1. Palmar view of the cleft hand with absence of the three central digits

Fig. 2. a Flatness of the lateral chest wall and little elevation of the right nipple. **b** With right arm elevated – Some fibers of the clavicular part of pectoralis major muscle are seen

Fig. 3. Absent middle phalanx of the little finger and hypoplasia of the phalanges and metacarpals of the three central digits



the thumb had a full range of active extension and flexion. Pinch and opposition was not possible due to poor movement and a lack of enough web space between the thumb and little finger. Chest examination showed flattening of the right anterior chest wall, absence of both the sternocostal part of pectoralis major and pectoralis minor muscle and a hypoplastic and elevated right nipple (Fig. 2a, b). Skiagram of the right hand showed absence of middle phalanx of the little finger. Remaining phalanges and metacarpals were hypoplastic (Fig. 3). Skiagram of the chest showed narrow intercostal spaces on the affected side.

On September 8, 1984 a Z-plasty was performed to deepen the web space and at operation it was further confirmed that the tendons of the flexor digitorum superficialis and profundus of the little finger were absent. There was no evidence of a rudimentary fibrous cord in place of the flexor tendons. On July 20, 1985 transposition of the little finger to the fourth metacarpal was performed. The finger was fixed in the functioning position with two small Kirschner wires. Eight weeks later the patient had satisfactory grasp and pinch action between the mobile thumb and the static little finger.

Discussion

The exact etiology of Poland's syndrome is not known. It does not appear to be genetically transmitted. It could be due to some kind of intrauterine insult at the seventh week of gestation when the developing limb bud with webbed fingers is adjacent to the chest wall. A recent report [12] has made an attempt to divide hand deformities associated with pectoral agenesis. *Type 1:* Five digits present even if hypoplastic. *Type 2:* Functional border digits with absent central digits. *Type 3:* More severe absence deformities with no functional digit. *Type 4:* Radial ray defect with absent thumb.

The case described here belongs to Type 2 group. Abnormalities of the tendons of the hand in patients of Poland's Syndrome are quite rare. But in our case the flexor tendons of the little finger were absent, this was confirmed on operative exploration. Ireland [8] reported one patient with absence of extensor tendons of all four fingers. Hiroaki [7] reported a case where the flexor tendons of the little finger in a case of Poland's Syndrome were thought to be absent clinically. On exploration they found that both tendons were adherent to each other and attached to the base of the proximal phalanx.

References

1. Beals RK, Crawford S (1976) Congenital absence of the pectoral muscle. A review of twenty-five patients. *Clin Orthop* 119:166–171
2. Brook Salap FS, Grauvier L (1971) Poland's syndrome. *Am J Dis Children* 121:263–264
3. Brown JB, McDowell F (1940) Syndactylism with absence of the pectoralis major. *Surgery* 7:599–601

4. Clarkson P (1962) Poland syndactyly. *Guy's Hosp Rep* 111:335–346
5. Epstein LI, Bennett JE (1970) Syndactyly with ipsilateral chest deformity. *Plast Reconstr Surg* 46:236–240
6. Goldberg MJ, Mazzei RJ (1977) Poland syndrome. A concept of pathogenesis based on limb bud embryology. *Birth Defects* 13:103
7. Hiroaki S, Tsuneichi E, Atushi H (1962) Anatomical findings in the hands of patients with Poland's syndrome. Report of four cases. *J Bone Joint Surg* 64A:1079
8. Ireland DCR, Rakayama N, Flatt AE (1976) Poland's syndrome. A review of forty-three cases. *J Bone Joint Surg* 58A:52–58
9. Mace JW, Kaplah JM, Schanberger JE, Cotlin RW (1972) Poland's syndrome. *Clin Paediatr* 11:98
10. McGillivray BC, Lowry RB (1977) Poland's syndrome in British Columbia. Incidence and reproductive experience of affected persons. *Am J Med Genet* 1:65
11. Poland A (1941) Deficiency of the pectoral muscles. *Guy's Hosp Rep* 6:191–198
12. Steven HG, Roy AM, Yoshio S (1984) Severe limb deficiency in Poland's syndrome. *Clin Orthop* 185:9
13. Walker JC, Meijer R, Aranda D (1969) Syndactylism with deformity of the pectoralis muscle. Poland's syndrome. *J Paediatr Surg* 4:569