POLAND SYNDROME : A CASE REPORT

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Introduction

Poland's syndrome has an incidence of 1 in 30,000, though other estimates vary from 1 : 1000 to 1 : 100,000 of all births (Chautard and Freira-Maia 1971). This consists of two main components, symbrachydactyly and pectoral muscle defect. Symbrachydactyly is a specific hand malformation, always unilateral, characterised by the association of short digits and syndactyly. The phalanges are short or absent. The middle phalanges are affected more frequently and in some cases they are obsent or fused with the distal phalnges. Syndactyly is either partial or complete, usually involving the soft tissue and not associated with bone synostosis. Syndactyly frequently involves the index and middle fingers. The associated muscle defect is an ipsilateral absence of sternal part of pectrol-is major and minor and maldevelopment of the external oblique and serratous anterior muscles. The associated defects include unilateral aplasia of kidney and hemivertebrae (Mace et al 1972). The incidence in this country is not described and we have not come across a report.

Case Report

A 6 year old male child was admitted on 3.8.77 to JIPMER Hospital with deformity of right upper limb since birth. He was the third child among four male children born to non consanguinous parents. There was no history of any congenital abnormality amongst siblings or close relatives. There was no history of injestion of drugs by the mother during pregnancy.

On examination, the child was moderately built and well nourished. There was flattening of the right side of the chest wall. The right nipple and areola were well developed and normal in position. On adduction of the arm against resistance, it was found that the right pectoral muscles were absent. The clavicular part of pectoralis major was well developed. Both scapulae appeared equal and normal in position (See Fig. 1 A).

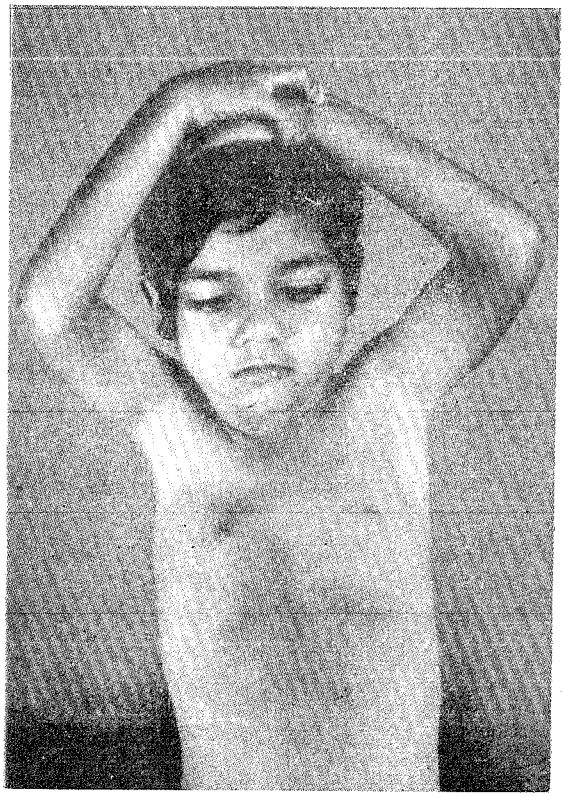

Fig. 1 A Note complete absence of Pectoral fold.

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The whole of the right upper limb was shorter by 2.5 cm. compared to the left. There was syndactyly of the right index and middle fingers. The right index was short and had only two phalanges. There was also a shallow web between the right middle and ring fingers. The web space between the thumb and index was also not fully formed, limiting free mobility of thumb. (Fig. 1 b)

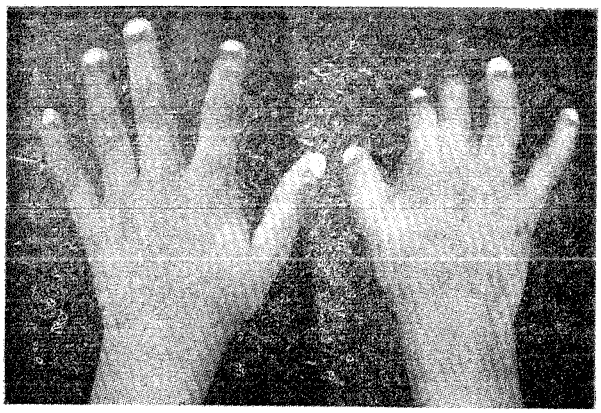

Fig. 1 B Deformity hand. There is partial syndaetyly of I, II & III webs and considerable Brachydactyly of Radial 3 digits.

Skiagrams of both hands were compared. The index and middle finger of right hand were smaller by 1.5 cms and the index had only two phalanges. The thumb was short. X-rays of scapualae and dorsolumbar spine were normal except for a ipsilateral small scapula. An I. V. P. was done, which showed normal sized kidneys with good excretion.

The patient was operated on 18.8.78 under general anaesthesia in a asanguinous field. The index and middle fingers were separated with zig-zag flaps and the cleft-deepened and covered with a dorsal tongue shaped flap, which was drawninto a nick made on the palmaer aspect, so as to avoid a bridling scar. The residual raw areas on the digits, which could not be covered by flaps, were grafted with full thickness grafts obtained from the ulnar border of the palm. The web space between middle and ring fingers was deepened using dorsal tongue shaped flap, interdigitating into a suitable nick on the palmer aspect (vide supra). The base of the digits was covered with skin flap based on palmer aspect. The cleft between the thumb and index was deepened using four flap 'Z' plasty.

Brock's type stirups anchored to a Cramer wire splint frame held the finger in the corrected position. The wounds were largely exposed except for the flaving wool packs on grafted areas. The wound healed well, and sutures were removed after 14 days.

Discussion

Though named after Alfred Poland, his description in 1841 made no references to the deformity of the hand and he was also not the first person to describe it (RAVITCH 1977).

Involvement of hand was referred to by FURST in 1900 but a detailed description was given only by Clarkson in 1962. As such this eponym is questionable (Mac Dowell 1977; Ravitch 1977).

Mace et al in 1972 collected 48 cases and added 7 cases of their own and they also brought out the association with ipsilateral hemivertebrae and renal aplasia. They found a male prepondlance of 3:1 and right side involvement in 75%. There was no evidence of hereditary transmisson. An intravenous pyelogram is recommended.

Summary

A case of Poland's syndrome characterised by anomalies in the shoulder girdle and symbrachydactyly is reported. First web was also involved. Method of correction adopted by us is described.

References

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