These devices illustrate some of the considerations used when designing for the developmental patterns of paediatric prostheses. Their design addresses developmental stages between the ages of four months and four years, not only by size changes, but also with the provision of a variety of control options.

REFERENCES

Poland's syndrome is an anomaly characterised by thoracic wall abnormalities, affecting the same side and deformities of the upper extremity. The thoracic wall deformity was first described by Lallemand in 1826. In 1841 Alfred Poland reported the post-mortem evaluation of a patient with deficient pectoralis major muscle and syndactyly of the hand on the ipsilateral side. The syndrome was named Poland's syndrome by Clarkson in 1962, after he had operated on a case similar to that described by Poland more than a century earlier.

The deformity is seen in 1 in every 25,000 births. It is sporadic in most cases, although familial patterns of inheritance have been reported. The ratio of male-to-female patients is 3:1 and, for unknown reasons, the deformity involves the right side in 75% of patients. The most common and classic presentation consist of simple syndactyly, limb hypoplasia and ipsilateral pectoral muscle agenesis. The degrees of limb deficiency have been classified into four groups by Gausewitz et al. Moderate to major limb wall deformities may need surgical reconstructions like iliacisthms, dorsal muscles transposition, autologous rib grafts, silicone implants have been reported. (Marks et al 1990).

Richard, aged 30, is a very interesting case of Poland's syndrome, a congenital problem and he was first seen at Nottingham Disablement Services Centre in 1992.

Richard was seen at Oxford Disablement Services Centre in 1975 when he was supplied with a partial hand leather palm case and gauntlet, having 4 rigid fingers and a thumb spring. He was also supplied with a working partial hand with split hook and tweezers. He became a good user with the split hook but disliked wearing it, being very self-conscious of his appearance and deformity. At the age of 17 he was advised to have a below elbow amputation so that a more functional and cosmetic prosthesis could be provided. Surgical intervention took place in June/July 1978 for the removal of the functionless right thumb and little finger.

In 1992 Richard was transferred from Oxford Disablement Services Centre to Leicester due to the redéfinition of Health Authority areas. On being seen at the local Disablement Services Centre it became apparent that Richard was not happy with the cosmetics and would like to be considered for a myoelectric prosthesis. As a result the Consultant referred him to the Trent Region Myo Team.

Medical Examination

Poland's syndrome affected Richard in the following manner: On examination, he had a two centimetre shortening of his right humerus and nine centimetre shortening of the right forearm with a partial palm with no fingers. Twenty five degrees of wrist flexion and extension were present, there was no pronation or supination.


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The right elbow joint had full flexion and extension, though there was marked varus deformity of his right elbow. Examination of the chest and shoulder revealed clinical absence of the radio ulnar joints.

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References


Authors

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Discussion

The provision of myoelectric prosthesis has resulted in complete reversal of his previous introvert and shy character to a much more relaxed and happy personality and using the prosthesis 7 days a week and up to 16 hours per day. He now uses his right hand functionally in almost everything as a physicist and is now engaged in research activities. Diagnosing his condition has allowed him to come to terms with his condition for the first time.

Management of the upper limb deficiency in Poland's syndrome with prostheses will probably only be necessary in patients belonging to types 3 and 4 because of more severe functional and cosmetic deficiencies. However, before surgery is contemplated especially in severely affected patients with severe deficiencies the opinion of an experienced rehabilitation team should be sought.

To our knowledge there has been no other reported cases of Poland's syndrome supplied with a myoelectric prosthesis.