PROVISION OF MYOELECTRIC PROSTHESIS FOR A POLAND'S SYNDROME PATIENT

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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[1]. 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[2]. 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[3].

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[4]. 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[5]. Moderate to major chest wall deformities may need surgical reconstructions like latissimus dorsi 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.

Prosthetic Background

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 re-definement of Health Authority areas. On being seen at the local Disablement Services Centre it became apparent that Richard was not happy with the cosmesis 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.

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 sterno costal head of his right pectoralis major. X-rays revealed fusion of the proximal radio ulnar joints with dysplastic changes in the distal radio ulnar joints. Several carpal bones were absent with presence of rudimentary metacarpals. The clinical features were typical of Poland's syndrome.

Myo Assessment

A detailed profile of the patient was built up by the clinical team (authors). This included past history, family details, social background and employment details. A complicated pattern emerged as although he trained as a physicist his occupation was a farm manager on his parents farm. He was also a lay-preacher in the local church. So his range of "employment" activities demanded a wide spectrum of prosthetic requirements from total cosmesis when conducting church services to manual duties on the farm. The most important being his concern of the cosmetic appearance of his prosthesis. The electromyographic signals obtained in the extensor and flexor muscles in the forearm were very good giving a good strong signal for the hand opening and closing functions respectively. It was then decided to proceed with a myoelectric prosthesis and a range of prosthetic hardware was shown to the patient so he could take part in the selection process. To be sure of maximum compliance from the patient this was thought to be a necessary and vital part of the treatment. The prescription of the prosthesis was an Otto Bock hand for a wrist disarticulation to enable us to keep the overall length as short as possible. The Otto Bock hand used has a non-detachable wrist unit for the minimum forearm overall length. In the past the prosthesis that had been previously manufactured were felt by the patient to be bulky and wide particularly at the wrist section.

Socket Construction

The socket is a two part design, the main anterior section consisting of hand, electrodes and battery. The posterior section being fitted after donning the anterior and held together by two velcro fastenings above and below the battery. The cup socket followed closely the contours of the patients hand. The socket was cut away around the patients very small little finger on the distal anterior surface allowing the finger to protrude through. (The main objections in the past being he thought the limb was too wide). The extensor section was contoured to fit over the elctrodes and give a neat appearance. The use of the removable section from the socket allowed easier application, removal and self suspension.

Training

At the delivery of the prosthesis a two day training programme started. This allows the patient once control has been established to be put into problem solving situations and to use the prosthesis in a natural way. Rapport was built up during the training period and there was an interchange of ideas between staff and patient. A suggested time plan was developed between therapist and patient of how to build up weight tolerance and a review appointment was given. At the return appointment the diary of compliance that the patient had kept over the previous month gave some surprising results when times of up to 16 hours per day of use had been recorded. A further checklist of timed task tests was recorded and this was repeated again after an interval of 4 months. The results seemed to indicated that bi-lateral skill remained fairly static but exclusive myo only tasks had improved.

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 and does not hide it in his pocket or behind his back as before. He has returned to the university as a physicist and is now engaged in research activities. Diagnosing his condition has made it easier for genetic counselling and also appropriate prosthetic provision 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 prosthetics will probably be only necessary in patients belonging to types 3 and 4 because of more severe functional and cosmetic deficiencies. However, before surgery is contemplated especially in patients with severe deficiencies the opinion of an experienced rehabilitation team should be requested. We would like to emphasise the importance of full examination and history taking of patients with upper limb deficiencies so that a correct diagnosis can be made at the earliest possible opportunity when formulating the prescription for a prosthesis and rehabilitation programme.

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

References

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