PROSTHETIC REHABILITATION OF BILATERAL ANTECUBITAL PTERYGIA WITH CONCOMITANT CONGENITAL HAND DEFICIENCIES: A CASE STUDY

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ABSTRACT

This case study presents upon the unique application of prosthetic rehabilitation principles for a young man presenting with bilateral antecubital pterygia or webbing of the elbows. His case was further complicated by substantial bilateral congenital hand deficiencies. Prior to prosthetic intervention, this patient's upper limb function was confined to midline manipulation performed with his elbows. Following a surgical release of the webbing of his right elbow he had sufficient mobility to justify an exploration of prosthetic rehabilitation. We report on the initial prosthetic fitting which has substantially expanded this young man's working envelop and upper limb function. This was accomplished through dual-site direct myolectric control of an electric hand mounted in relative internal rotation to facilitate midline function and preserve the patient's sensory input from his right residual forearm and hand.

INTRODUCTION

Antecubital pterygium syndrome has been defined as an extremely rare genetic disorder characterized by bilateral, fairly symmetric antecubital webbing extending from the distal third of the humerus to the proximal third of the forearm with associated musculoskeletal abnormalities. In the case in question, this physical presentation was further complicated by the presence of concomitant congenital hand deficiencies. Collectively, this left the patient largely bereft of meaningful upper limb function. This case describes the initial prosthetic fitting process and the subsequent improvements in upper limb functionality. Written informed consent was obtained for the presentation of this case study.

PATIENT PRESENTATION

The patient initially presented with bilateral webbing of the elbows restricting him to a non-functional 5-10° of elbow function (Figure 1). Distal to the elbow webbing, the patient presented with shortened forearms with significant congenital hand deficiencies inclusive of a single residual digit on his left upper extremity and two residual digits on his right extremity. Active wrist flexion and extension was present in both upper limbs (Figure 1,2). The patient's presentation was otherwise unremarkable with normal lower limb and cognitive function.

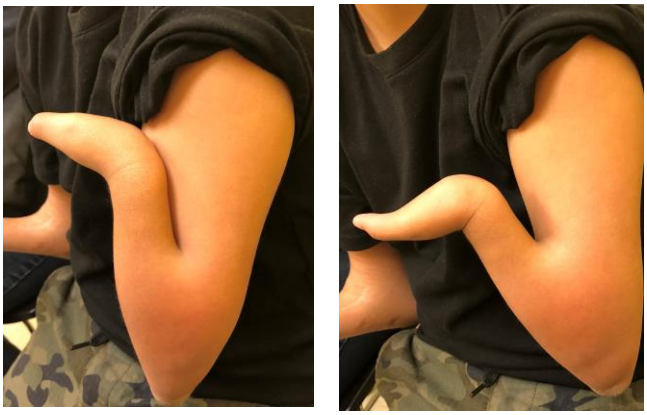

Figure 1: Left elbow and wrist mobility demonstrated in pictures of end range wrist and elbow flexion and end range wrist and elbow extension.

Following surgical release to the right elbow, the patient became capable of an expanded range of elbow motion, creating a possibility of additional functional gains through prosthetic rehabilitation (Figure 2).

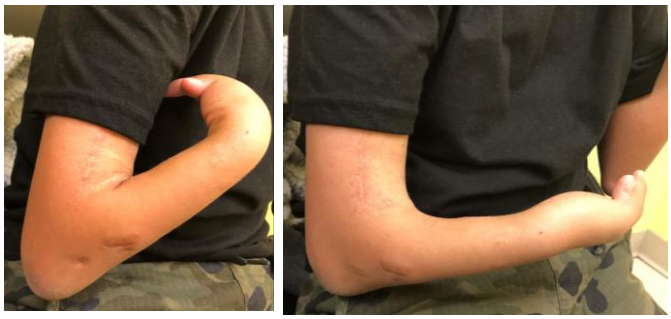

Figure 2: Right elbow and wrist mobility demonstrated in pictures of end range of wrist and elbow flexion and end range of wrist and elbow extension

Prior to prosthetic intervention, the patient's upper limb function was largely confined to bilateral manipulations performed by the elbows, or using his residual hands to

stabilize objects against his face to facilitate fine motor control. Writing, for example, was performed through neck movement with the residual limb stabilizing a pen against the head. Eating required the stabilization of a fork handle between the residual limb and head to stab a portion of food, followed by laying the fork on the table top where it could be bitten off of the fork.

PROSTHETIC MANAGEMENT

Recognizing the value of preserving the sensory input provided by the residual right forearm and hand, and the functional value of a mobile, sensate limb segment, the decision was made not to enclose the distal aspect of the limb within the socket. Further, recognizing the value of maintaining an anatomic length the prosthesis, the decision was made to mount the forearm, wrist and hand of the prosthesis in relative internal rotation. This was assessed dynamically in a test socket fitting prior to the completion of a definitive device (Figure 3).

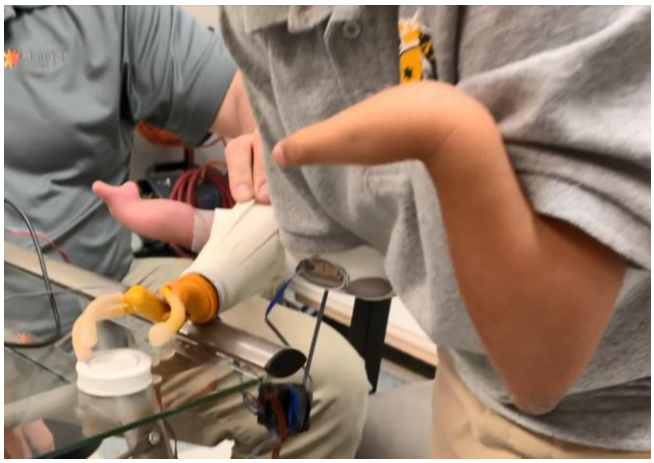

Figure 3: Dynamic assessment of the internally rotated prosthetic forearm to ensure optimal upper limb function.

Dual-site direct control was used to control prehension of a pediatric hand. EMG signals were obtained from the wrist flexors and extensors mounted within a costum silicone socket. The prosthesis also included an internal battery with an adjustable friction rotation wrist (Figure 4).

We subsequently observed that he needed increased wrist motion for different activities and installed a ball-in-socket universal friction wrist (myolino wrist 2000) to further increase his functional envelope. This increased his capacity to feed himself as well as write in a more ergonomic position.

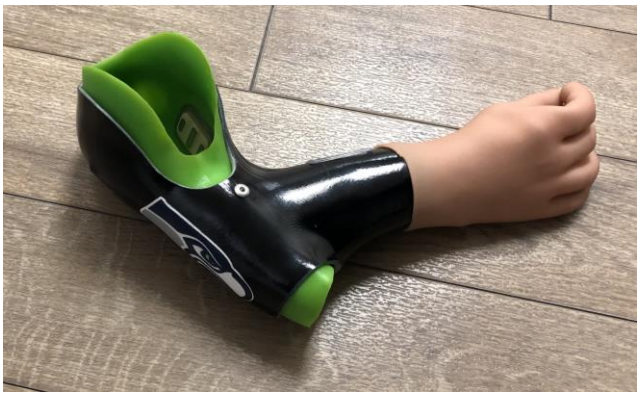

Figure 4: Definitvie dual-site myoelectric prosthesis

The functional benefits associated with the prosthesis were immediately apparent as the child demonstrated the ability to grasp and lift objects from a table. He is now able to write with the aide of the prosthesis, sitting erect with no need to lower his face to the table top. Similar benefits have been observed with eating as this young man is able to grasp a fork and raise it to his mouth with no need to lower his face to the table top. The prosthesis has broadly enabled other midline functions (Figure 5).

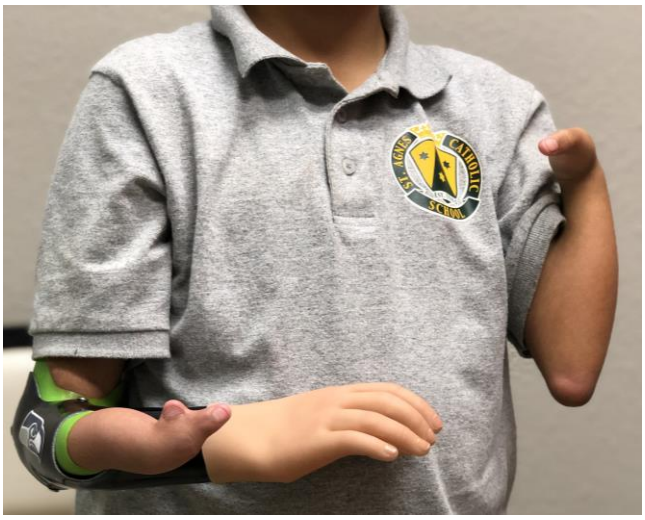

Figure 5: Definitive prosthesis fully donned, demonstrating the capacity for midline function.

Future prosthetic plans include replacing the current terminal device to a more durable small adult hand. The surgical team is not currently considering a soft-tissue release on the left extremity due to concerns of possible secondary damage to the neurovascular bundle. We have considered the construction of a pass-through elbow-disarticulation style prosthesis on the left to facilitate bimanual function such as personal hygiene, riding a bike or sports activities, but this has not yet been formally explored.

CONCLUSION

This case illustrates a novel application of prosthetic rehabilitation principles. The case demonstrates a careful balance between providing a prosthetic enhancement to the affected extremity without overly compromising its native movement and sensory input. While the resulting device is less physiologic in its appearance, it meets the predominant needs of the patient in allowing him to write and eat in a more acceptable body alignment and position.