

## Introduction to the Medical Abstracts and Annotated Bibliography

The abstracts and annotated bibliography present examples of my ability to read and synthesize information about advanced medical topics.

I chose to write about a rare genetic defect called Popliteal Pterygium Syndrome. This defect is usually passed from parent to child through autosomal dominant expression. Sometimes, however, there are spontaneous mutations, and the syndrome appears in a family with no other incidence of the defects related to the syndrome. The syndrome manifests itself through a long list of physical deformities, all of which are repairable to some degree through plastic and reconstructive surgery. The articles I chose discussed orthopedic treatment plans to correct the leg defects associated with the syndrome.

These works were for use by orthopedic surgeons, and were written to educate them about the syndrome and to assist them in deciding if the articles discussed therein are good sources of information. After reading the abstracts and/or bibliographies, the surgeons will read the articles I am abstracting and use the information presented in the articles to prepare an appropriate surgical treatment for this rare disorder.

I arranged the bibliographies in alphabetical order. The abstracts, I placed in publication date order, based on the assumption that my readers would be most interested in recently published findings and would then go back and look at the older literature.

I chose the most recently published articles that would allow my reader to compare/contrast a variety of surgical approaches to the syndrome. I tried to keep in mind the purpose of my communication when deciding what to include. Working on the assumption that surgeons would be looking for information to help determine a plan for surgery, I tried to drill down to that information and synthesize it into a useful, brief communication.

## **Medical Abstracts**

**Parikh, S.N.; Crawford, A.H. Do, T.T; Roy, D.R (2004). Popliteal Pterygium Syndrome: implications for orthopaedic management [Leg and Foot]. *Journal of Pediatric Orthopaedics, B*; 13(3), 197-201.**

Popliteal Pterygium Syndrome is a rare disorder, generally inherited as an autosomal dominant genetic trait, although sporadic cases are reported. This syndrome presents in approximately 1 of 300,000 live births. Less than 100 cases have been reported in the literature since 1968. This disorder manifests with a popliteal web and can include craniofacial and genitourinary malformations. Management of the fixed flexion deformity caused by the popliteal web present in most PPS cases is of utmost concern to the orthopedic surgeon. When determining appropriate treatment, the anatomy of the popliteal web must be known. Leg involvement can present in varying severity from simply a pigmented streak extending from upper thigh to heel to a web containing a dense, fibrous band running from the ischium to the calcaneus. In these latter cases, the sciatic nerve generally runs deep into the band and the hamstring and calf muscles can have anomalous insertion. Indications for surgical intervention include fixed flexion deformity of the knee and limitation in patient's ability to ambulate. Based on experience and literature review, two treatment protocols are recommended. When popliteal webbing is minimal, soft tissue release and multiple Z-plasties are recommended. A more aggressive approach is advocated when webbing is moderate to severe, including initial soft tissue release, followed by femoral shortening or supracondylar femoral extension osteotomy. Surgical intervention should begin early with multiple surgeries likely necessary. If the PPS presents with additional deformities requiring surgery, combination of surgeries should be considered to lessen patient stress.

**Gardetto, A and Piza-Katzer, H. (2003). A case of familial Popliteal Pterygium Syndrome: early surgical intervention for successful treatment. *Pediatric Surgery International*; 19(8), 612-14.**

A two-year-old boy presents with Popliteal Pterygium Syndrome (PPS) causing severely reduced range of motion in right knee and ankle. Surgery was undertaken to release the webbing to help accomplish knee extension, free the sciatic nerve and lengthen the Achilles tendon. Inspection after initial incision revealed that the sciatic nerve, located in the free edge of the popliteal web, was the limiting factor to full extension of the knee joint. Multiple Z-plasties and postoperative casting of leg in the full-extended position allowed some stretching of the nerve. Three months postoperatively, patient presented with full nerve function in both lower limbs. When flexion contracture of knee joint is greater than 45°, splinting and traction as a treatment for the flexion deformities are of limited success, therefore surgical intervention is becoming the treatment modality of choice. Early surgical intervention and appropriate procedures for dealing with the lengthening of the shortened sciatic nerve, including either nerve expansion or serial casting for periods greater than three months, can result in release of the flexion deformity and help increase patient's mobility and range of motion.

**Maral, T; Tuncali, D; Özgür F; Safak, T; and Gürsu, K.G. (1997). A case of Popliteal Pterygium Syndrome treated along with nerve expansion [case reports]. *Plastic and Reconstructive Surgery*; 100(1), 91-95**

Popliteal Pterygium Syndrome is a rare genetic syndrome. The most common manifestation of the syndrome is a web like skin formation in the popliteal area limiting full extension of the knee joint. Surgical management of cases becomes difficult when the sciatic nerve is involved. Z-plasties alone have proved to be insufficient and alternate traction and corrective splinting do not produce results. The case of a three-year-old boy for whom an unsuccessful Z-plasty had been undertaken is presented. In this case, the location of the sciatic nerve in the free edge of the web is the main hindrance to full extension of the knee. A 75cc rectangular tissue expander was inserted under the nerve, and filled with 30cc of saline. Expansion of the nerve began three weeks postoperatively, with 5cc of saline injected weekly. After six weeks, wound dehiscence was noted and the procedure terminated, however measurements indicated a 60% ratio of nerve elongation. Extension of the knee joint measured at 160° compared to 120° preoperatively, showing a 40° gain in extension. Although it is possible the common peroneal division of the sciatic nerve may be susceptible to limited injury, nerve expansion and repair are advantageous for three reasons: 1) The intact nerve is not harmed; 2) Diameter discrepancy is avoided; and 3) The regenerating axons pass over only one coaptation site. With use of electromyography or measurement of the intraluminal pressure of the tissue expander, some adverse affects can be avoided. Though treatment in this particular case terminated early and there is a lack of additional patients, nerve expansion has good potential as an alternative treatment modality for patients with popliteal webbing.

## Annotated Bibliography

**Gardetto, A and Piza-Katzer, H. (2003). A case of familial Popliteal Pterygium Syndrome: early surgical intervention for successful treatment. *Pediatric Surgery International*; 19(8), 612-14.**

Surgeons from the department of Plastic and Reconstructive Surgery at Innsbruck University Hospital, Innsbruck, Austria discuss the use of Z-plasty to release extensive skin webbing, and lengthen the sciatic nerve and Achilles tendon of a two-year-old patient presenting with Popliteal Pterygium Syndrome. Their findings indicate that the shortened sciatic nerve causes the greatest challenge to repair. Early surgical intervention coupled with slow postoperative stretching of the sciatic nerve is the appropriate treatment for patients with knee contractures greater than 45°. This is a similar plan to that advocated by Parikh, et.al., although the approach is more conservative.

**Maral, T; Tuncali, D; Özgür F; Safak, T; and Gürsu, K.G. (1997). A case of Popliteal Pterygium Syndrome treated along with nerve expansion [case reports]. *Plastic and Reconstructive Surgery*; 100(1), 91-95**

Surgeons from the Department of Plastic and Reconstructive Surgery at Hacettepe University Faculty of Medicine, Ankara, Turkey discuss lengthening the sciatic nerve in patients with Popliteal Pterygium Syndrome. The shortened nerve limits full knee extension in these patients. Following a Z-plasty soft tissue release, a procedure advocated by Gardetto, et.al., and Parikh, et.al; a 75-cc tissue expander is inserted and injected with 5 cc of saline weekly, to attempt gradual elongation of the nerve. After 6 weeks, the procedure was terminated, yet the authors note a 60% ratio of nerve elongation; an improvement over the average 40% increase noted by Miller.

**Parikh, S.N.; Crawford, A.H. Do, T.T; Roy, D.R (2004). Popliteal Pterygium Syndrome: implications for orthopaedic management [Leg and Foot]. *Journal of Pediatric Orthopaedics, B*; 13(3), 197-201.**

Surgeons with the division of Pediatric Orthopaedic Surgery at Children's Hospital Medical Center, Cincinnati, Ohio discuss surgical strategies for management of the fixed flexion deformities often associated with Popliteal Pterygium Syndrome. This group of surgeons has handled eight cases of the disorder since 1978, a large number considering the rarity of Popliteal Pterygium Syndrome, which occurs in approximately 1:300,000 live births. Two different approaches are advocated, based on the severity of the deformity. For severe cases, an aggressive strategy including either femoral shortening or femoral extension osteotomy is recommended. Z-plasty with soft tissue release is recommended in cases with lesser involvement.