

Case Study: Vincenzo

Conditions Treated

Arthrogryposis with Lower Limb Deformities,
Severe Contractures, & Pterygium

Age Range During Treatment

14 years to 16 years

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BACKGROUND

Vincenzo was a 14 year old boy from Bari, Italy who was diagnosed with arthrogryposis at birth and had been wheelchair bound since the age of six. His family had previously sought treatment in Altamura and Milano Italy but doctors had not provided any treatment and told the family that nothing could be done to improve his condition. Vincenzo had a burning desire to walk and wanted to have better control of and movement in his upper limbs. The family traveled to America for definitive care.



A photo of Vincenzo during an early office visit.

EVALUATION

- Vincenzo's lower extremities demonstrated active hip flexion and extension of 4/5 strength. There were 20 degree hip flexion contractures with the ability to flex the hip to 90 degrees.
- X-rays revealed severe knee flexion contractures with skin webbing (pterygium) on the right. The left knee ranged from 45 to 95 degrees of flexion and the right knee ranged from 85 to 110 degrees flexion. Active flexion and extension indicated that he had enough strength to overcome gravity.



X-rays of Vincenzo's right and left knees, tibias, and fibulas.

- He was unable to walk more than one step.
- His left foot had a severe fixed equinovalgus deformity, the right ankle / foot was not as severely valgus but had a fixed 30 degree equinovalgus deformity along with a talar dome irregularity.



Vincenzo's feet showing equinovalgus deformities.

- Vincenzo's shoulders had atrophied leaving him unable to reach forward and above his head but I was able to move his arms (passively) 70 degrees forward and overhead.
- His triceps functioned 4/5 but he had no bicep function. The left elbow was fixed in full extension, and the right elbow had a range of motion of 0 to 20 degrees. X-rays revealed elbow irregularities and minimal joint space.



Vincenzo's right elbow.

- His left wrist was in approximately 80 degrees of flexion with minimal grasp strength and could be passively extended to about 50 degrees of flexion. The right wrist was in 60 degrees of flexion and could be extended passively to 30 degrees.
- His finger grasp function was weak on the right but better than the left and he was able to write with his right hand by maneuvering his body.

TREATMENT

I determined that surgery was possible as a result of Vincenzo's age and the complexity of his contractures. With the assistance of two interpreters I explained the surgical plan along with the risks and potential benefits to Vincenzo and his mother and answered all of their questions in detail.

Surgery

- **Left Knee Posterior Release Including Capsule & Peroneal Medial Nerve Release**
- **Left Foot Osteotomies & Soft Tissue Releases**
- **Right Foot Soft Tissue Release**

Left Knee Posterior Release Including Capsule & Peroneal Medial Nerve Release

An incision was made over the upper part of the knee (distal knee) and incisions were made into the hamstrings and calf (gastrocnemius) allowing the muscles to be lengthened (release). The knee joint's capsule was opened to expose the neurovascular bundle at which point the peroneal nerve was released all the way down into the anterior compartment of the leg (neurolysis) and the posterior capsule was incised. At this point, it became possible to bring the knee from an approximately 60 degree flexion contracture to the normal position of being fully straight.

Left Foot Osteotomies & Soft Tissue Releases

Incisions were made over the calcaneus and talonavicular joints. The foot was preoperatively dislocated at the talonavicular joint and the calcaneocuboid joint was shifted in an outward direction to properly position the ankle. At this point tenotomies were performed on the peroneus longus and brevis. An osteotomy was performed on the talonavicular joint to achieve dorsiflexion of the foot to neutral position and was held in place with pins. The calcaneus joint was opened (bones were repositioned) and held in place with a large bone graft.

Right Foot Soft Tissue Release

A tenotomy was performed on the Achilles tendon and the posterior capsule was opened, allowing the foot to be brought into a neutral position.

The wounds were irrigated and closed in layers. Bilateral short leg casts were applied and a knee immobilizer was applied to the left knee.

Observations

Vincenzo's bilateral lower limb casts were removed a month after surgery, by which time his left knee's range of motion had increased from 45-90 degrees to 0-90 degrees.

Surgery

- **Right Posterior Knee Release Include Hamstrings, Gastrocnemius, Soft Tissue Fascia, Fasciectomy, Neurovascular Bundle Neurolysis, Posterior Capsulotomy For Right Knee Release**
- **Patellar Relocation of Chronically Dislocated Right Patella**
- **Application of External Fixator w/ Ilizarov For Hinged Internal Fixation**

Right Posterior Knee Release Include Hamstrings, Gastrocnemius, Soft Tissue Fascia, Fasciectomy, Neurovascular Bundle Neurolysis, Posterior Capsulotomy For Right Knee Release

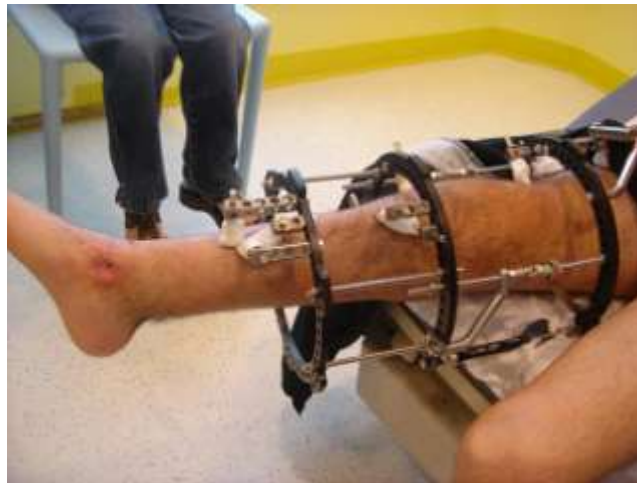
A large posterior incision was made and the fascia was released around the sciatic nerve as well as the neurovascular nerve. The hamstring tendons were also released. The capsule was released and a neurolysis was performed on the peroneal nerve as well. His knee was then brought from an approximately 90 degree knee flexion contracture to approximately 60 degrees. A Z-plasty of the posterior skin was performed by Dr. Michael Margiotta of NYU Plastic Surgery.

Patellar Relocation of Chronically Dislocated Right Patella

The right knee had a chronically dislocated knee cap (patella) so a quadricepsplasty was performed. The patella was located by extending the leg and shifting the patella into proper position.

Application of External Fixator w/ Ilizarov For Hinged Internal Fixation

Closures of the surgical incisions were performed by Dr. Margiotta. As the wounds were closed, an Ilizarov fixator was applied to the femur with two hinges at the knee along with wires and pins at the tibia to achieve good fixation and correction.



Ilizarov external fixator applied to right leg.

Observations

- Following his second surgery, the range of motion in Vincenzo's left knee was at 0-75 degrees and both feet were properly touching the ground (plantigrade). He continued taking antibiotics and using a continuous passive motion machine (CPM) for his left knee along with distraction of the right knee. Vincenzo wore a foot plate attached to his Ilirazov frame for positioning along with a knee brace and ankle foot orthoses (AFO).
- Vincenzo's foot healed properly. I also stressed the importance of range of motion exercises for his left knee and right foot and that they were to continue advancing his knee frame 1 mm per day. He was fitted for a new AFO which appeared to fit perfectly and was only to be removed for bathing.

- 3 weeks after surgery, Vincenzo had his sutures removed by Dr. Margiotta. In collaboration with Dr. Margiotta, it was decided that the knee extension be slowed down with a three day pause followed by Vincenzo's mother advancing his frame by 1 mm per day. His knees were straightening and his feet were still plantigrade.
- Within a few months, the quads and hamstrings in Vincenzo's left leg became active and he was able to move his left knee from 0 to 90 degrees. The flexion contracture in his right knee decreased and he was eventually able to move his right knee from 0 to 45 degrees.

Surgery

Removal of Hardware

Removal of Hardware

All hardware, pins, and wires were removed from Vincenzo's legs. X-rays showed good positioning of his knees. He was fitted for bilateral leg braces and then had bilateral Bledsoe braces applied to his knees.



Observations

- 6 days after having his hardware removed, Vincenzo was able to walk from the waiting room in my office to an exam room with assistance.
- Within two and a half weeks, Vincenzo was walking at a faster pace and his mother discontinued use of his wheelchair. His surgical sites were fully healed and he had good range of motion in his knees. He was fitted for new braces that were to be worn with his knees in an alternating locked and unlocked position.
- A month later Vincenzo came into the office for a final follow-up visit before his return to Italy. Vincenzo was now able to walk independently and x-rays showed good healing and positioning of his knees. Plans were made for him to continue diligent physical therapy with his mother as well as aquatic therapy.

Surgery

**Right Elbow Release With Radial Head Excision And
Ulnar Nerve Transposition**

Right Elbow Release With Radial Head Excision And Ulnar Nerve Transposition

A posterior incision was made over the elbow, the ulnar nerve was moved forward (transposed anteriorly) and the intermuscular septum was cut. The elbow's joint capsule was opened laterally and the anterior capsule was removed in its entirety along with the posterior capsule. At this point, the elbow was able to be moved from 0 to 40 degrees.

The anterior structures were protected but there was still impingement of the radial head and the triceps tendon was quite tight. A V-Y plasty of triceps tendon was performed.

The patient now went from 0 to 60 degrees. At this point, the radial head was noted to be impinging and was excised. After excision of the radial head, the joint went from about 0 to almost 90 or 100 degrees. However, the olecranon was noted to be hinging rather than gliding smoothly. It was felt that an outer bridge was needed. The elbow now had a range of 0-110 degrees and we felt that it would be terrific if we could keep this motion. The ulnar nerve was then transposed anteriorly and held in place with a soft tissue sleeve.

Post Op

- Vincenzo's mother began range of motion exercises for his elbow 48 hours after surgery and a continuous passive movement (CPM) machine was ordered.
- Two weeks after surgery, Vincenzo had 0 to 90 degrees of range of motion in his right elbow and was able to move his fingers. He was to continue his exercises and use of the CPM machine to encourage greater movement ability and use of his hands and fingers.

CONCLUSION

It's been eight years since Vincenzo's last surgery and his legs are doing very well and while he is still wearing braces, he is able to walk without assistance.

His upper extremities have maintained passive motion which allows him to utilize his hand in ways he could not before. To date we have not performed any additional procedures to achieve active motion.



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