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Orthopedic Surgery Considerations in Post-Polio Syndrome

Poliomyelitis vaccination has made acute polio infection a rarity in the United States and most developed nations. Now, 40 to 50 years after initial infection, polio survivors are experiencing the effects of post–polio syndrome. With symptoms such as worsening muscle weakness, muscle pain, severe fatigue, and new-onset weakness, they are constantly seeking care from orthopedic surgeons. Conservative therapies, ranging from activity modification, range-of-motion exercises, and orthotic and brace support to surgery, provide pain relief and help to optimize extremity function. With the potential for functional decline secondary to muscle overuse, patients with post–polio syndrome should be closely monitored, and treatments should be implemented at appropriate times to maximize function and ensure patients' independence.

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Poliomyelitis is a disease that is caused by an enterovirus and that initially presents with gastrointestinal symptoms. As the virus gains access to the central nervous system and attacks the anterior horn cells of the spinal cord, the disease process may present with minor symptoms, such as pain and stiffness after a febrile illness, or more severe symptoms, such as paralysis. The last major US epidemic occurred in the early 1950s. Since then, effective immunization programs have made acute poliomyelitis a rarity in the United States and other developed nations. The paralysis resulting from polio remains a lifelong challenge, however, as survivors experience musculoskeletal limitations that may eventually worsen and lead to complete disability and loss of independence. This major health problem continues to require the expertise of orthopedic surgeons, who must treat patients with post–poliomyelitis syndrome.

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Poliomyelitis Classification and Post–Polio Syndrome

Poliomyelitis is classified into 4 distinct categories based temporally on the onset of infection onset. During the **acute phase** of the infection, 95% of the anterior horn cells, which control the skeletal muscle cells of the trunk and limbs, are attacked, accounting for the diffuse and severe paralysis typically observed. With bulbar involvement, paralysis of the respiratory muscles can be life-threatening and may require mechanical ventilation. Respiratory compromise should be suspected with shoulder involvement because of the close proximity of the anterior horn cells controlling each anatomic region within the spinal cord. A variable number of anterior horn cells survive the initial infection. Anterior horn cells either undergo cell death within 3 days or recover over 1 month.

In the **subacute phase** of the disease, anterior horn cell survival, axon sprouting, and muscle hypertrophy occur and represent the 3 mechanisms by which patients regain strength. A mean of 47% (range, 12%-94%) of the anterior horn cells in the spinal cord survives the initial attack. The pattern of cell survival occurs randomly. Final distribution of muscle paralysis is inconsistent between different patients and depends on which anterior horn cells have undergone irreversible destruction. Each anterior horn cell is responsible for innervating a specific group of muscle cells. When a group of muscle cells is "orphaned" by the death of the anterior horn cell that

Name | Eval Date | DOB | Age

Height | Weight | Lbs | Right Hand: Pinch | Kgs | Grip | Kgs

Pulse | BP | Left Hand: Pinch | Kgs | Grip | Kgs

MANUAL MUSCLE TESTING AND JOINT EXAMINATION:

Neck ° | Flexion ROM | ° | Extension RO M | Neck Strength | Flexion | Extension

Scoliosis | Pelvis Obliquity | SI Tenderness: | Sciatic Notch Tenderness

UPPER EXTREMITY:	RT	LT
SHOULDER		
Shrug		
Flexion		
Abduction		
Extension		
Adduction		
Internal Rotation		
External Rotation		
Adson's Test		
Biceps Tenderness		
Supraspinatus Tenderness		
Pain with Restricted Abduction		
Palpable Cuff Defect		
A-C Joint Tenderness		
ELBOW		
Flexion		
Extension		
Pronation		
Supination		
Med Epicondyle Tenderness		
Lat Epicondyle Tenderness		
Pain with Wrist Extension		
Pain with Wrist Flexion		
WRIST HAND		
Wrist Flexion		
Wrist Extension		
Finger Flexion		
Finger Extension		
Thumb Flexion		
Thumb Extension		
Thumb Opposition		
Thumb Adduction		
Thumb Abduction		
Median Tinel's Sign		
Phalen's Test		
Thenar Atrop hy		
Thumb CMC Deformity		
Comments:		

LOWER EXTREMITY:	RT	LT
HIP		
Range of Motion (Ext -Flex)		
Flexion		
Extens ion		
Abduction		
Adduction		
Trendelenberg Sign		
Trochanteric Tenderness		
KNEE		
Range of Motion (Ext -Flex)		
Extension		
Flexion		
Genu Varum		
Genu Valgum		
Genu Recurvatum		
Joint Effusion		
Patello -Femoral Crepitation		
Ilio -Tibia l Band Contractures		
LEG AND FOOT		
Ankle Range of Motion (PF -DF)		
Tibialis Anterior		
Extensor Hallucis		
Extensor Digit orum		
Peroneal		
Flexor Digitorum		
Flexor Hallucis		
Tibialis Posterior		
Gastroc -Soleus		
# Single Leg Toe Raises		
Foot Deformity		
Plantar Fascia Tenderness		
LEG MEASUREMENTS		
Actual Leg Length ASIS		
Apparent Leg Length (Umbilicus)		
Thigh Circumference		
Calf Circumference		
GAIT		
Aids Used		
Description		

POLIO EVALUATION - Continued

DIAGNOSIS	
Post Polio Syndrome	138.0
Unequal Leg Length	736.81
Abnormality of Gait	781.2
Leg Muscle Paresis	344.30
Cervical Spondylosis	721.0
Cervical Stenosis	723.0
Lumbar Spondylosis	721.3
Lumbar Stenosis	724.02
Kyphosis	737.10
Scoliosis	737.30
Thoracic Outlet Syndrome	353.0
Shoulder Impingement	726.2
Biceps Tendinitis	726.12
Supraspinatus tendonitis	840.6
Rotator Cuff Tear	727.61
Shoulder Osteoarthrosis	715.91
Lateral Epicondylitis	726.32
Medial Epicondylitis	726.31
Elbow Osteoarthrosis	715.92
Trigger finger	727.03
Wrist Osteoarthrosis	705.93
Hand Osteoarthrosis	715.94
DeQuervain's Tenosynovitis	727.04
Pelvic Obliquity	738.6
Hip Osteoarthrosis	715.95
Trochanteric Tendinitis	726.5
Hip Subluxation	718.25
Chondromalacia Patella	717.7
Genu Valgum	736.41
Genu Varum	736.42
Genu Recurvatum	736.5
Kne e Osteoarthrosis	715.96
Ilio -Tibial Band Contracture	718.46
Equinus	736.72
Foot Planovalgus	754.69
Foot Cavus	736.73
Foot Cavovarus	736.75
Foot Equinovarus	736.71
Foot/Ankle Valgus	736.79
Foot/Ankle Osteoarthrosis	715.97
Plantar Fasci itis	728.71
Hallux Valgus	735.0
Hallux Rigidus	735.2

RECOMMENDATIONS :

Lifestyle modification to avoid muscle overuse

Weight loss program

More frequent rest periods to avoid fatigue

Modified exercise program with minimal resistiv e exercises and frequent rest periods

Stretching exercises

Breathing exercise program

Use of elevated seating to avoid shoulder strain

Braces AFO Right Left

KAFO Right Left

Spine:

Use of UE walking aids: | Cane Crutches

Wheelchair/Seatin g Clinic Referral

Gait Laboratory evaluation

Handicapped parking privileges

Use of power mobility devices: Electric Scooter | Electric Wheelchair Stair Chair Glide | Power lift seat on chair

Change in Working Status

Sleep Apnea Study

Oth er Testing:

Surgery

Medications

Referrals

Other

Polio Clinic Follow - up Appointment

Figure 1. Sample physical examination form for assessment of polio survivors. (Copyright held by Dr. Keenan.)

supports it, nearby nerve cells may sprout additional axons and "adopt" some of the orphaned cells. The process of axon sprouting allows further expansion of a motor unit (defined as a nerve cell and the muscle cells it innervates). In addition, compensatory enlargement or hypertrophy of muscle cells within a specific unit provides a supplementary method by which patient strength is enhanced.

The **residual poliomyelitis phase** is the period 16 to 24 months after initial infection. During this period, it is possible to determine the ultimate extent and residual deficits of poliomyelitis. During this time, procedures to restore lost function and grant structural stability can first be used.

A forth distinct phase has been recognized in patients who had acute poliomyelitis during childhood and who demonstrate **worsening or new onset of weakness 30 to 40 years after initial infection**. This weakness is not a result of infectious spread of the earlier disease but, rather, is caused by overuse of the muscles that were originally affected, whether or not they were known to have been affected at disease onset. Studies have shown that a muscle must lose from 30% to 40% of its strength for weakness to be detected using manual muscle testing. Gait studies have also demonstrated that activities of daily living require more muscle strength and stamina than previously appreciated. The traditional protocol, which encourages patients to work harder to regain strength, was proved to be detrimental and to lead to chronic overuse of muscles and further functional decline.

The diagnosis of post–polio syndrome is made with a positive history for poliomyelitis and associated symptoms, including increased muscle weakness of a variable pattern, muscle pain, severe fatigue, muscle cramping or fasciculations, joint pain or instability, sleep apnea, intolerance to cold, and depression. Pathognomonic tests for the syndrome are not available. Electromyography can demonstrate the presence of large motor units resulting from the previous axon sprouting, but this finding is supportive and not diagnostic of poliomyelitis.

Patient Evaluation

Polio survivors require a thorough musculoskeletal evaluation consisting of a detailed history and physical examination. After the initial evaluation, these patients should be reevaluated annually for functional deterioration.

It is imperative to obtain information regarding the acute infection phase and to determine the patient's functional level beyond the initial infection into the recovery phase. Important components of the history include age at initial infection, muscle groups and extremities primarily involved, pulmonary involvement, requirement of mechanical ventilatory support, degree of disability and exhibition of independence, history of falls, use of braces, ability to ambulate, and worsening or new symptoms.

The physical examination requires a comprehensive documentation of range of motion (ROM), manual muscle testing, and presence of deformity for each portion of the axial skeleton and for each joint. Specialized tests, such grip testing and finger pinch, should be conducted, along with others that assist in defining common orthopedic manifestations of post–polio syndrome (Figure 1).

Treatment Approach

In the acute phase of the disease process, the most important consideration is to assess the patient for bulbar involvement and the need for mechanical ventilation. Pulmonary compromise can be fatal in the acute phase without proper supportive treatment. Other measures are aimed at decreasing muscle pain and preventing future complications. Instituting regular ROM exercises can assist in preventing joint contractures.

During the subacute phase, which may last as long as 24 months, the emphasis is on preventing deformities and preserving function. Splints and brace devices may be helpful in maintaining joint position and enhancing existing function.

Patients who exhibit compromised function of the diaphragm as a result of bulbar involvement can be taught glossopharyngeal breathing. This technique, in which air is swallowed into the lungs, provides sufficient air exchange for the patient to perform light activities in the sitting position. Mechanical support of ventilation may still be required while the patient sleeps. It is during this residual stage that orthopedic surgery is commonly performed to restore lost function and provide structural stability. In the skeletally immature patient, it is important to prevent skeletal deformities resulting from muscle imbalance. Before any surgery requiring general anesthesia, vital capacity must be documented to determine the patient's need for respiratory support.

Patients with post–polio syndrome benefit from treatment protocols directed at preserving current muscle strength and preventing further weakness. In general, restoration of strength to a muscle that has been weakened by poliomyelitis is not typical. However, some gain in muscle strength can be expected when chronic overuse is corrected through activity and behavior modification. Instituting a limited exercise program with minimal resistance and frequent rest periods is useful in preventing muscle disuse atrophy and weakness.

Orthoses and Bracing

Lightweight orthotic support of the limbs is useful in protecting joint function. For the lower extremity, orthoses assist mainly in the stance phase of gait; they substitute for weak muscles by way of mechanical realignments (Figure 2). An example of this is limiting ankle dorsiflexion to neutral when there is weakness of the plantar flexor muscles. As the body's center of mass moves forward, the ankle plantar flexors contract to control the forward motion of the tibia over the foot. If the ankle plantar flexor muscles are too weak to control the tibia, then limiting dorsiflexion range will provide control of tibial motion. The overall goal of these devices is to maintain joint alignment and to limit active force generation across the joint. Orthoses are a crucial adjunct to patient education and activity modification in treating patients with post-polio syndrome.

Surgery

Orthopedic surgery is occasionally recommended in the treatment of polio survivors. The goals of surgery include pain relief, deformity correction, muscle transfers to counter muscle force imbalance, stabilizing unstable joints, and allowing for more effective use of orthoses and braces. Surgery does not eliminate the need to use braces and does not improve muscle strength. Specific management strategies depend on the areas of disease involvement.

Upper Extremity

Shoulder. Shoulder weakness is found in 95% of patients with post–polio syndrome and correlates closely with degree of lower extremity weakness. The shoulder joint, which is completely dependent on muscle strength for active mobility, is important for placement of the hand in its desired position for use. In patients with post-polio syndrome, preservation of shoulder strength should be a treatment priority, especially in ambulatory patients who require upper extremity aids. Whether shoulder function is preserved is one factor determining a patient's independence.

Lower extremity weakness and disability lead to excessive demands on the shoulders. Patients with weak lower extremities use their arms to raise themselves from seats, pull themselves up stairs, and transfer themselves in and out of cars. They also lean heavily on upper extremity aids while ambulating. Therefore, it is important to remove as many unnecessary strains on the shoulders as possible. This can be done with elevated seats, motorized lift chairs, elevators or motorized stair chair glides, and optimal lower extremity bracing. With increased loading of the shoulder, however, rotator cuff tears are a common problem; surgical repair should be done when possible. For large tears not amenable to repair, arthroscopic débridement offers significant pain relief.

In patients who require a wheelchair, weak muscles about the shoulder girdle can be made more functional with use of mobile arm supports on the wheelchair. These supports allow for a larger arc of motion and are advantageous for patients with less muscle strength. A glenohumeral arthrodesis may benefit the patient with sufficient strength in the scapulothoracic muscles. When the shoulder is fused, scapulothoracic motion is maintained, allowing use of the extremity for tabletop activities. Glenohumeral fusion restricts the patient's ability to position the hand for bathroom hygiene, so performing the procedure on both shoulders is undesirable.

In minimally ambulatory or nonambulatory patients, an electric wheelchair or motorized scooter should be prescribed to prevent excessive strain on the shoulder muscles caused by propelling a manual wheelchair.

Wrist/Hand. Wrist involvement is common, as several patients with post–polio syndrome use crutches or canes as assists for ambulation. The chronic pressure placed on the wrist leads to an increased incidence of carpal tunnel syndrome, often requiring surgical release of the deep volar ligament. These patients also experience carpal

LOWER LIMB ORTHOTIC PRESCRIPTION

Name: **Date:**

Diagnosis: **Duration of Use:** ☐ Temporary ☐ Permanent

Prognosis: ☐ Excellent ☐ Good ☐ Fair ☐ Poor

Brace Type: ☐ UCBL ☐ Arizona ☐ Knee Cage ☐ AFO ☐ Rear Entry, Floor Reaction AFO ☐ KAFO ☐ HKAFO ☐ Other:

Side: ☐ Right ☐ Left ☐ Bilateral

Construction: ☐ Polypropylene ☐ Copolymer ☐ Thermoplastic Elastic ☐ Metal and Leather

Closures: ☐ Velcro ☐ Laces ☐ Buckles ☐ Elastic ☐ Right Hand Pull ☐ Left Hand Pull

Ankle Design:

☐ Non-Articulated

Resistance to Dorsiflexion: ☐ PLS ☐ Minimum ☐ Moderate ☐ Maximum

☐ Articulated

Type of Joint: ☐ Tamerak ☐ Wafer ☐ Integrated ☐ Lawrence Other:

ANKLE MOTION: ☐ None ☐ Range of Motion : Dorsiflexion: ° Plantarflexion:

☐ Spring Dorsiflexion Assistance ☐ 90 Degree Dorsiflexion Stop

FOOT PLATE: ☐ _ Length Longitudianl Arch ☐ Long Toe Plate ☐ Padded ☐ Metatarsal Pad

KNEE JOINT: ☐ Single Axis ☐ Posterior Offset ☐ Polycentric ☐ Dynamic Knee Extension

KNEE LOCK: ☐ None ☐ Drop Locks ☐ Bail Locks ☐ Spring Loaded Other:

KNEE CONTROL:

☐ Varus ☐ Valgus ☐ Hyperextension ☐ Infrapatellar Strap ☐ Anterior Knee Pad

THIGH SECTION: ☐ Quadrilateral ☐ Ischial Weight Bearing ☐ Extended Lateral Wall

HIP CONTROL: ☐ Silesian Belt ☐ External Hip Joint

HIP MOTION: ☐ Abduction: ° ☐ Flexion: ° ☐ Extension: °

SHOES: ☐ Canvas ☐ Oxford ☐ Orthopaedic ☐ Extra-depth ☐ High Top ☐ Molded

OTHER: ☐ Ankle Strap ☐ Posterior Strap to Limit Dorsiflexion

SPECIAL INSTRUCTIONS:

Figure 2. Sample lower extremity orthotic prescription form domonstrating available bracing options. (Copyright held by Dr. Keenan.)

bone subluxation as well as wrist arthritis secondary to the increased forces transmitted across the joint. Joint arthrodesis, an effective treatment to relieve pain and restore stability to the wrist joint, may be required.

Almost 50% of hand function loss may occur with opponens paralysis, and hand function loss is common in patients with post-polio syndrome. A splint placed during the acute and recovery phases is useful in preventing adduction contractures, whereas tendon transfers made at the appropriate time may restore opponens function. The most common muscle transferred is the flexor digitorum superficialis of the ring finger.

Paralysis of the intrinsic muscles of the hand interferes with function. A lumbrical bar orthosis will prevent hyperextension of the metacarpophalangeal joints and allow the long extensors to extend the fingers and open the hand. Surgical capsulodesis and related procedures to limit metacarpophalangeal joint extension provide the same result.

When paralysis is present, finger flexor and extensor action can be supplied by a flexor hinge orthosis, if wrist extensor function is present. Tendon transfers can provide the same result, allowing the tenodesis effect to provide grasp and pinch functions.

Lower Extremity

Hip. Full hip and knee ROM is imperative for proper lower extremity function and an energy-efficient gait. Contractures must be corrected (when possible) to permit more effective bracing. Total hip arthroplasty (THA) has proved to be a useful addition to the armamentarium for treating patients with post-polio syndrome and hip involvement. Because of contractures about the hip and muscle force imbalance, these patients are predisposed to hip arthritis. Factors to consider in identifying the proper THA candidate include presence of flexion contractures, degree of joint laxity, muscle strength, adequate bone stock in the case of osteoporosis, and the patient's ability to adhere to postoperative limitations and rehabilitation protocols. Excessive hip muscle weakness is a contraindication to THA. The patient must exhibit a minimum of grade 3 hip abductor, flexor, and extensor strength, and there must be other lower extremity deformities requiring correction before prosthetic replacement can be considered. Surgery can be expected to weaken the surrounding muscles; to prevent chronic dislocation, this result must be taken into account before THA is undertaken.

A series of 500 patients with postpoliomyelitis were followed prospectively by Dr. Keenan in a specialized clinic (Hosalkar H, Fuller DA, Kay D, Esquenazi A, Israelite C, Keenan MA. Functional outcomes of lower extremity total joint arthroplasty in patients with post-polio syndrome. Presented at the Annual Meeting of the American Academy of Orthopaedic Surgeons, February, 2007, San Diego, CA. *Proceedings*. 2007;3:652). Over a 15-year period, there were 9 candidates for THA for the treatment of osteoarthritis. These 9 patients underwent THA, and their Harris Hip Scores improved from 94 (range, 62-111) to 173 (range, 162-182). This increase was statistically significant ($P<.05$). Four hips had preoperative flexion contractures (mean, 33°; range, 15°-45°). All flexion contractures were corrected at time of surgery, with no postoperative recurrence or instability (Figure 3).

Knee. The typical knee deformity in post–polio syndrome is valgus alignment. This deformity can be secondary to iliotibial band contracture, hip adduction contracture, leg length discrepancy (short leg), or valgus foot.

Iliotibial band contractures are common deformities. The hip assumes a position of flexion, external rotation, and abduction while the knee exhibits valgus alignment and the tibia is externally rotated on the femur. Surgical release or lengthening of the iliotibial band will correct the deformity for both joints. Complete release should be avoided, as the iliotibial band is a supporting structure during the stance phase of gait.

A varus deformity of the knee is also possible. This deformity is characteristically associated with osteoarthritis. If the deformity is flexible, a knee–ankle–foot orthosis (KAFO) may be effective, but it is less so than for valgus deformities. In severe osteoarthritic knees, total knee arthroplasty (TKA) should be considered.

A patient with flail lower extremities can stand with crutch assistance and a KAFO with the knees locked in extension and the ankles in slight dorsiflexion by hyperextending the hips and using the robust anterior hip capsule for support. Flexion contractures of the hips or knees prevent this alignment. If trunk support and upper extremity strength are adequate, the

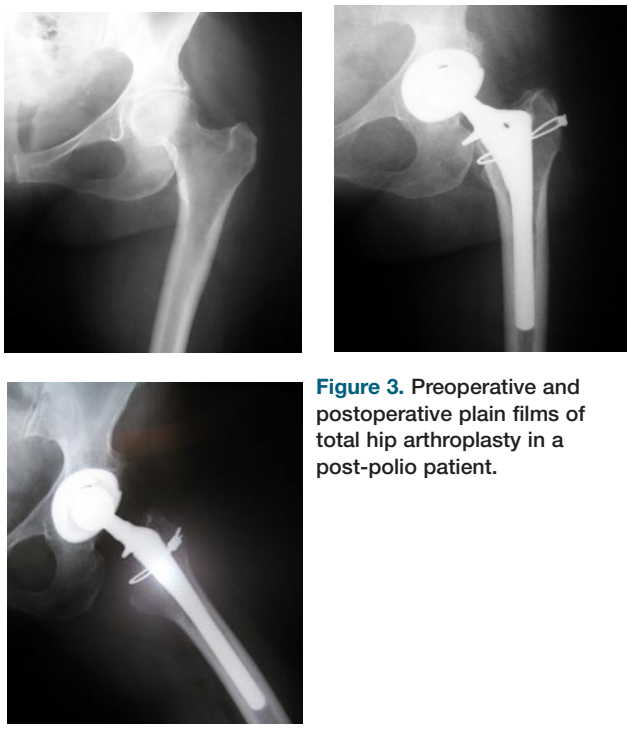

Figure 3. Preoperative and postoperative plain films of total hip arthroplasty in a post-polio patient.

Figure 4. Preoperative and postoperative films of total knee arthroplasty in a post-polio patient.

patient could ambulate with a swing-through gait for short distances, though this gait has high energy demands.

Over time, the posterior knee joint capsule becomes stretched, and this knee laxity leads to a painful recurvatum deformity and arthritic degeneration of the knee. A KAFO will protect the knee and provide improved stability for walking, specifically in patients with more severe recurvatum. Patients with mild deformity may benefit from a short leg brace ankle–foot orthosis. As reconstruction of the posterior knee capsule is not feasible, TKA is a possible treatment option, especially in patients with concomitant arthritis.

If there is fair (grade 3) strength in the hip flexor muscles and passive full-knee extension, then the knee joints can be left unlocked for walking. In this case, a posteriorly offset knee joint is used to stabilize the knee, and ankle dorsiflexion is limited to -3° of neutral dorsiflexion to provide a hyperextension moment to the knee for stability. Thus, at stance phase, the net ankle plantarflexion locks the knee in hyperextension, restrained by posterior capsular static structures.

Flexion contractures of the knee require constant use of the quadriceps muscle throughout the gait cycle—resulting in excessive fatigue, which is difficult for most patients to tolerate. Surgical release of the hamstrings, as well as posterior knee capsulectomy, can be effective in treating the problem. Ultimately, without treatment, the ability to ambulate decreases, and patients are left predisposed to arthritis. TKA in this situation is a treatment option that can be considered.

From the above-mentioned study by Hosalkar and colleagues, 10 patients underwent TKA. Knee Society Scores improved from 28 (range, 0-49) to 88 (range, 85-93). This increase was statistically significant ($P<.01$). Five knees had preoperative valgus deformity (mean, 13°; range, 11°-17°), and 1 had mild varus deformity (14°). For all patients before surgery, the mechanical axis passed through the center of the knee in the coronal plane. Preoperatively, 3 knees had a recurvatum deformity (10°, 25°, 50°), and 1 had a flexion contracture (10°). Postoperatively, 2 knees had a flexion contracture (10°, 15°), and the other 8 achieved full extension with no recurvatum (Figure 4).

Foot/Ankle. Muscle imbalances in the foot can lead to deformity. Tendon releases or transfers should be considered before these imbalances develop into fixed deformities.

Equinus contracture of the ankle is a common problem and results in genu recurvatum. Accommodating the equinus posture by using an elevated heel places excessive stress on the calf muscles to control the leg. Surgery to lengthen the Achilles tendon is often needed to correct the equinus contracture of the ankle and to permit adequate bracing (Figure 5).

A cavus foot deformity causes forefoot equinus, which also limits bracing. In the absence of bony abnormalities, release of the plantar fascia is sufficient to correct the deformity. If the cavus deformity is caused by bony abnormalities, then a closing wedge osteotomy through the cuneiform bones is required. A triple arthrodesis of the hindfoot can also be used to correct deformities and provide a stable base of support.

Conclusions

Poliomyelitis vaccination has made acute polio infection a rarity in the United States and most developed nations. Now, 40 to 50 years after initial infection, polio survivors are experiencing the effects of post–polio syndrome. With symptoms such as worsening muscle weakness, muscle pain, severe fatigue, and new-onset weakness, they are constantly seeking care from orthopedic surgeons. Conservative

Figure 5. Illustration of Hoke triple hemisection technique for Achilles tendon lengthening.

therapies, ranging from activity modification, ROM exercises, and orthotic and brace support to surgery, provide pain relief and help to optimize extremity function. With the potential for functional decline secondary to muscle overuse, patients with post–polio syndrome should be closely monitored, and treatments should be implemented at appropriate times to maximize function and ensure patient independence.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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