NEUROMUSCULAR CONDITIONS

Poliomyelitis: Orthopaedic management

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Summary
Although poliomyelitis has been eradicated in most parts of the world, orthopaedic surgeons occasionally encounter residual deformities in patients who suffered the disease in childhood. An understanding of the causative factors and the available treatment options are essential before surgical intervention is contemplated. It is also well recognised that post-polio syndrome occurs in those who suffered poliomyelitis 20–40 years ago.

It is important to note that inadequate or improper surgical intervention can potentially lead to more disability; a well-planned approach to the particular part of the body affected by polio, after considering the patient as a whole and understanding the principles involved, is the best option.

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History of poliomyelitis
Poliomyelitis is said to have first occurred nearly 6000 years ago in the time of the Ancient Egyptians. The evidence for this is in the withered and deformed limbs of certain Egyptian mummies. The following are the more important dates in the history of polio (Fig. 1):

Ancient Egypt 3700 BC: An Egyptian mummy with probable polio. If this was polio, cases almost certainly occurred before then.

1580–1350 BC: The Priest Ruma with a withered leg and equinus foot—shown on a plaque and probably poliomyelitis.

1209 BC: Mummy Giptah with an equinus foot.

(Middle Ages) 1559: A painting by Pieter Bruegel shows a crippled beggar. This is not definitely polio, although it probably did occur during this period in England.

(Eighteenth century) 1789: First known description of poliomyelitis by Underwood.

(Nineteenth century) 1834: First epidemic of poliomyelitis in the island of St. Helena.

1855: First description by Duchenne of the pathological process in poliomyelitis involving the anterior horn cells of the spinal cord.

(Twentieth century) 1908: Transmission of poliomyelitis to a monkey by Landsteiner.

1909: Passage of the virus through a monkey by Flexner.

1949: Growth of the virus on tissue culture.

1951: Three types of polio virus isolated and identified.

1954: First large-scale trial of Salk (live attenuated vaccine) by injection.

1958: First general use of Sabin (live attenuated vaccine) by mouth.
Epidemiology

Since the World Health Assembly resolved in May 1988 to eradicate poliomyelitis, the estimated global incidence of polio has decreased by more than 99% and three World Health Organization (WHO) regions (Americas, Western Pacific, and European) have been certified as polio-free. Since 1994, when the countries of the WHO South-East Asia Region (SEAR) began accelerating polio-eradication activities, substantial progress towards that goal has been made. By 2001, poliovirus circulation in India had been limited primarily to the two northern states of Uttar Pradesh and Bihar, with 268 cases reported nationwide. However, a major resurgence of polio occurred during 2002, with 1600 cases detected in India, of which 1363 (85%) were in Uttar Pradesh and Bihar. Problems remain due to the difficulties involved in extending immunization coverage to some regions (especially Africa), integrating new vaccines into routine immunization schedules and securing sufficient funding for programs. Injection safety is also a major problem that should be resolved by utilisation and proper disposal of single-use auto-disabling syringes. The forthcoming availability of new vaccines and the action of the Global Alliance for Vaccines and Immunization hold reasonable hope for the future. Other problems remain, such as new conditions resembling polio paralysis caused by viral infection other than by poliovirus and post-polio syndrome (PPS).

It is safe to assume that acute and residual poliomyelitis is still unfortunately encountered in the developing world. In developed countries, on the other hand, residual poliomyelitis is still occasionally seen in the elderly and immigrants.

Acute poliomyelitis

Poliovirus is primarily spread by faecal–hand–oral transmission from one host to another. The virus is shed in oral secretions for several weeks and in the faeces for several months. It destroys the anterior horn cells in the spinal cord.

Poliavirus infections can be divided into minor and major forms:

1. The minor illnesses occur 1–3 days before the onset of paralysis, with gastrointestinal complaints such as nausea and vomiting, abdominal cramps and pain, and diarrhoea. There are also systemic manifestations such as sore throat, fever, malaise, and headache. This stage lasts usually for 2–3 weeks but, may extend for up to 2 months; the presence of any tenderness in the muscles is evidence that the acute stage is not over. The major illness includes all forms of central nervous system (CNS) disease caused by poliovirus, including aseptic meningitis (or non-paralytic polio), polio encephalitis, bulbar polio, and paralytic poliomyelitis, alone or in combination.

The clinical findings associated with an attack of polio are as follows:

1. There is fever, stiffness of the neck (nuchal rigidity), and a pleocytosis in cerebrospinal fluid.
2. Profound asymmetrical muscle weakness develops.
3. The initial phase is typically followed by some recovery of muscle strength, but permanent weakness results from necrosis of anterior horn cells.
4. Rarely, a transverse myelitis with paraparesis, urinary retention, sensory symptoms and signs along with autonomic dysfunction (including hyperhidrosis or hypohidrosis), and decreased limb temperature may occur.

In this stage, the treatment is mainly medical involving the paediatric physicians. General supportive treatment for the pyrexia and irritation, the prevention of secondary respiratory infection and the treatment of any respiratory paralysis are the main aspects of the treatment.

The paralysed legs are supported by plaster splints or pillows and sandbags to keep the hip joints in 5° of flexion and in neutral rotation. The knee joint is held at 5° of flexion and the foot is supported in a 90° position. Splinting relieves pain and spasm and prevents the development of deformities.

Recovery stage

In this stage, also known as the convalescent stage, the acute symptoms and muscle tenderness disappear and the paralysed muscles begin to recover. This stage lasts for up to 2 years after the onset of the disease. During this whole period, there is gradual recovery of the muscles; the recovery is rapid in the first 6 months but is slower during the subsequent months.
Treatment in this stage is mainly in the orthopaedic department involving physiotherapy and splinting. The aims of treatment are:

(a) To assist in the recovery of paralysed muscles by remedial exercises.
(b) To prevent deformities by the use of orthotic appliances.

An assessment is first made of the extent of muscle paralysis by charting the power of various groups of muscles and grading them according to the international nomenclature (Medical Research Council grading) as follows:

0 — Complete paralysis
1 — Slight flicker of contraction present
2 — Muscle can move a joint only when gravity is eliminated.
3 — Muscle can move a joint against gravity and resistance
5 — Full normal power.

Total functional assessment of the limbs is made before planning treatment. This will include:

(a) Charting the muscle power grades.
(b) Extent of contractures and deformities.
(c) Method of ambulation.
(d) Shortening of the limb.

Efficient physiotherapy is the mainstay of the management of this stage of poliomyelitis. A good physiotherapy department with facilities for exercise therapy, hydrotherapy and electrical stimulation of muscles is essential in the management of paralytic polio.

Orthotic management

Appropriate orthotic appliances are prescribed to prevent deformities due to muscle imbalance, as shown in the following chart.

The new international nomenclature for orthotic appliances describes the joints that are stabilised by the appliance e.g. the name 'Ankle Foot Orthosis' replaces the old term 'below knee appliances'.

When the power of muscles controlling the hip and knee are normal and the weakness is only in the dorsiflexors or plantar flexors of the ankle or invertors or evertors of the foot, the patient is prescribed an Ankle Foot Orthosis (Below Knee orthosis or Caliper).

Indications for orthotic appliances

<table>
<thead>
<tr>
<th>Muscle paralysed</th>
<th>Deformity</th>
<th>Appliances</th>
</tr>
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<tbody>
<tr>
<td>Foot Evertors</td>
<td>Inversion</td>
<td>Ankle Foot Orthosis (AFO) with inside bar and Outside T strap</td>
</tr>
<tr>
<td>Invertors</td>
<td>Eversion of foot</td>
<td>Ankle Foot Orthosis (AFO) with outside bar and Inside T strap</td>
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When the quadriceps power is 2 or below, the knee has to be stabilized and hence a Knee Ankle Foot Orthosis (Full or Above Knee Calliper) is prescribed.

If the hip abduction power is poor i.e. less than, 2, the appliance will include a pelvic band with a hip joint (Hip Ankle Foot Orthosis) to prevent the lurching gluteus medius gait.

In the recovery stage, a child who starts with a full appliance with pelvic band may be able to gradually manage with shorter appliance and ultimately discard it when the muscles fully recover due to intensive physiotherapy treatment.

Residual paralysis stage

The period beyond 2 years after the onset of the disease is called the residual paralysis stage. No recovery of muscle power occurs in this stage. Deformities are liable to occur due to imbalance of muscle power and poor posture. There is also disuse atrophy of muscles and shortening of the leg due to interference with growth. In neglected cases, gross fixed deformities of the hip, knee and foot occur with severe wasting of muscles. Children with extensive paralysis and gross deformities have to crawl on all fours to move from place to place.

Post-polio syndrome

PPS is the term used for the newly occurring late manifestations of poliomyelitis that develop in patients 30 to 40 years after the occurrence of the acute illness. It has been estimated that 25–60% of the patients who had acute polio may experience these late effects of the disease. The specific cause of post polio syndrome is unknown; the aetiology has been attributed to pathophysiological and functional causes. Pathophysiologic causes include chronic poliovirus infection, death of the remaining motor neurons with ageing, premature ageing, damage to the remaining motor neurons caused by increased demands or secondary insults, and immune-mediated syndromes. Functional
aetiologies for PPS include greater energy expenditure as a result of weight gain and muscle weakness caused by overuse or disuse. PPS has been recognized for over 100 years, but is more common at the present time because of the large epidemics of poliomyelitis that occurred in the 1940s and 1950s. PPS is characterised by neurological, musculoskeletal, and general manifestations. Musculoskeletal manifestations include muscle pain, joint pain, spinal spondylolisthesis and scoliosis, and secondary root and peripheral nerve compression. General manifestations include generalised fatigue and cold intolerance. The slowly progressive muscle weakness occurs in those muscle groups already involved such as the quadriceps and calf muscles.

Diagnostic criteria for post-polio syndrome

1. A prior episode of paralytic poliomyelitis with residual motor neuron loss (which can be confirmed through a typical patient history, a neurological examination, and, if needed, an electrodiagnostic exam).
2. A period of neurological recovery followed by an interval (usually 15 years or more) of neurological and functional stability.
3. A gradual or abrupt onset of new weakness or abnormal muscle fatigue (decreased endurance), muscle atrophy, or generalized fatigue.
4. Exclusion of medical, orthopaedic, and neurological conditions that may be causing the symptoms mentioned in 3.

Management of post-polio syndrome

Many patients require revision of orthotic devices such as braces, canes, and crutches or to use new lighter orthotic devices to treat new symptoms. Common issues include genu recurvatum, knee pain, back pain, degenerative arthritis, or arthralgia. Surgery for scoliosis or fractures may also be necessary to treat new conditions.

Pattern of muscle weakness and deformities

Upper limb involvement

Late functional deterioration is common in long-term poliomyelitis patients. While upper-limb pain in individual functional regions is common, its overall prevalence and pattern in long-term poliomyelitis is poorly documented. There are data in support of ‘overuse’ due to greater mobility aid dependence as a cause of increasing upper limb pain in long-term poliomyelitis especially among severely paralysed polio patients.

Lower limb involvement

Typical osseous or soft-tissue abnormalities about the knees affected by poliomyelitis include external rotation of the tibia, excessive valgus alignment, ligamentous laxity, and genu recurvatum.

With localised wasting, the quadriceps can help compensate for a weak calf. With hamstring weakness the ability to decelerate the tibia is lost and, therefore, flexion of the knee will persist throughout the stance phase. In order to prevent this the patient may attempt to compensate with increased quadriceps activity for a longer portion of the stance phase. In the case of a weak quadriceps and hamstrings the occurrence of an equinus contracture, or a hinged AFO with a dorsiflexion block will both prevent excessive knee flexion and excessive ankle dorsiflexion during the stance phase.

The pitfall of lengthening of the Achilles tendon should be avoided in these patients. These patients may require an ischial bearing, double upright locked knee orthosis, which helps prevent the knee from buckling during gait.

Common foot and ankle deformities seen are pes cavovarus (hindfoot cavus) due to evertor paralysis (peroneus brevis and longus) and pronated everted foot due to invertor paralysis (tibialis anterior and posterior). Foot intrinsics are typically spared in polio. Claw toes result from relative overactivity of the long toe flexors and extensors (to compensate for weakness of the triceps)

Mortality

Excluding polio patients with respiratory failure, long-term mortality following polio appears to increase 20 years after recovery from the acute illness. Contracting severe paralytic poliomyelitis at a young age seems to increase long-term mortality.

Treatment

It is more economic to prevent 100 polio cases than to treat one hopelessly crippled child. It is often quicker to straighten 100 deformed limbs by simple subcutaneous surgery than to treat a single patient with complicated procedures. The final aim should be that patients return to their own village or town, accepted and integrated into their own communities, and earning their own living among their own friends.

Since overuse weakness is frequently present in these patients, the role of slowly progressive non-fatiguing exercise in their rehabilitation is emphasised. New muscle weakness of a mild-to-moderate degree responds well to a non-fatiguing exercise program and pacing of activity with rest periods to avoid muscle overuse. Generalised fatigue may be treated with energy conservation and weight loss programs and lower extremity orthoses.

An orthosis is a device which externally supports an existing body part with the objective of supporting, correcting or compensating for skeletal deformity or weakness. The current range of available orthoses is many and varied and with the advent of new materials such as carbon fibre, advanced manufacturing techniques and the range of devices available to the prescriber is ever increasing. Orthoses are available for all parts of body and aid in conservative and definitive treatment of many deformities. The thermoplastic leaf spring AFO, or drop foot splint, is one good example of an orthosis commonly used. It assists dorsiflexion and uses 3-point pressure to stabilise the ankle joint.
**Orthopaedic operations in patients with residual poliomyelitis**

**Hip and knee contractures of over 30°**

In general these will all require surgery, unless one or both arms are weak in addition to bilateral lower limb paralysis, when the use of crutches will be difficult or impossible. In a young child with fairly recent contractures the most important single factor responsible for the deformity is a tight tensor fascia lata and ilio-tibial band. In the older child or adult, however, other ligamentous and tendinous structures play an important part and must be divided as well.

The subcutaneous method of division is very satisfactory for less severe contractures, provided it is done correctly and as extensively as necessary. Care must be taken to avoid damaging the femoral and popliteal arteries and the common peroneal nerve. The biceps, however, should always be divided under direct vision because of the risk of damaging the adjacent lateral popliteal nerve.

**Tendon transfer to re-establish muscle power**

In selecting a tendon to transfer, the muscle should be sufficiently strong to supplement the power of a paralysed muscle. The nerve and blood supply of the transferred muscle should be preserved in order to avoid iatrogenic weakness.

For efficiency the transferred tendon should be securely attached (with tension) close to the insertion of a paralysed tendon, and should be routed in a direct line between its origin and the new insertion.

The transferred tendon should also be retained in its own sheath, avoiding tunnels in fascia, bone, or an interosseous membrane to avoid adhesions.

The joint across which the muscle acts must be in a satisfactory position; all contracted structures must be released before the tendon transfer.

When possible an agonist muscle, with the same range of excursion of its tendon, should be chosen.

**Muscle transplantation to replace a paralysed muscle**

In these procedures, unlike tendon transfer, both the origin and the insertion of a muscle are detached along with its neurovascular pedicle. This procedure is not as popular as tendon transfer, because of the difficulty in finding a normal muscle to transplant, donor side morbidity, the technical difficulty and the shortage of microvascular surgeons in the third world countries where residual polio is still seen.

**Stabilization of relaxed or flail joint**

Tenodesis, fixation of ligaments, and construction of artificial check ligaments are used to restrict the range of movement or to eliminate abnormal motion of a joint. With few exceptions, these procedures have been discarded, as deformity in the opposite direction may occur, and the tendon or artificial check ligaments may stretch with time.

This technique can still be helpful in skeletal immature patients.

**Arthrodesis**

Arthrodesis is used to correct deformity, relieve pain in arthritic joints and reduce the number of joints a weak muscle is acting across orthroclesis is more popular than tenodesis. In skeletal immature patients, extra-articular arthrodoses can be performed, allowing continued growth of the skeleton.

**Limb lengthening**

Often poliomyelitis is unilateral causing limb length inequality, which occasionally requires limb lengthening. In leg lengthening for patients with poliomyelitis, callus maturation is slow and patients tend to develop contractures despite physiotherapy, bracing or joint fixation. Concomitant and secondary surgery is frequently required to treat associated problems or residual deformities. Lengthening along an intramedullary locked nail can significantly shorten the treatment time with relatively few complications.

**Joint replacement surgery**

In patients with post-polio residual deformities joint replacement can be indicated. In one study, pain and knee scores improved following total knee arthroplasty in patients with a history of poliomyelitis and antigravity quadriceps strength, but there was less pain relief in patients with less than antigravity quadriceps strength. Recurrence of instability and progressive functional deterioration is possible in all knees affected by poliomyelitis that have undergone total knee replacement, but they appear to occur more commonly in more severely affected knees.

**Ilizarov techniques**

There are many drawbacks to using conventional approaches for the treatment of complex foot deformities, like the increased risk of neurovascular injury, soft-tissue injury, and shortening of the foot. An alternative approach, that can eliminate these problems is the Ilizarov method. Pin-tract problems, contractures, residual and recurrence of deformity can complicate the Ilizarov method.

**Conclusion**

The surgeon managing the residual weakness of poliomyelitis and post-polio syndrome must possesses an understanding of the pathological process in poliomyelitis as well as the variations in the pattern of the disease in different parts of the body. Poliomyelitis causes a lower motor neuron disease unlike other types of neuromuscular paralysis. The neurological problems, and the pattern of paralysis following poliomyelitis is different from upper motor neuron paralysis or indeed lower motor neuron paralysis caused by other diseases.
Knowledge of patho-anatomy before embarking on surgery is necessary. The mainstay of management remains physiotherapy and orthotic appliances.

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