PART II – NEUROLOGICAL DISORDERS

CHAPTER 10
PARAPLEGIA NON TRAUMATIC

Dr William P. Howlett
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CHAPTER 10

PARAPLEGIA NON TRAUMATIC

Introduction
Paraplegia (PP) means paralysis of the legs. It is caused mainly by disorders of the spinal cord and the cauda equina. They are classified as traumatic and non traumatic. Traumatic paraplegia occurs mostly as a result of road traffic accidents (RTA) and falls (Chapter 19). Non traumatic paraplegia (NTP) has multiple causes (Table 10.1) and is the most common cause of adult neurological hospital admissions in Africa after stroke and infection. Paraplegia is among the most common community based neurological disorders in Africa. The aim of this chapter is to review the causes and management of NTP. The student should aim to be able to localise the site and classify the main causes of paraplegia and to investigate and manage a patient presenting with it.

AETIOLOGY
Paraplegia can arise from a lesion either within or outside the spinal cord or cauda equina. These are classified as compressive and non compressive. Compression is caused either by bone or other masses. The main compressive causes are Pott’s disease (TB of spine) and tumours (usually metastases). The main non compressive causes are transverse myelitis secondary to viral infections, HIV, TB and very occasionally syphilis. Less common causes include Devic’s disease, B-12 deficiency and helminthic infections. There also exists in Africa a large group of nutrition related non compressive paraplegias, termed the tropical myeloneuropathies. These include konzo, lathyrism and tropical ataxic neuropathy (TAN). The main causes of paraplegia are presented in Table 10.1.

Table 10.1 Main causes of nontraumatic paraplegia in Africa

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Common causes of paraplegia in Africa

- Pott’s disease (TB)
- inflammation (transverse myelitis)
- malignancy (metastases)
- infection (HIV)
- nutritional (konzo)

Localization/anatomy
The spinal cord extends from C1 in the neck to the lower border of L1 (Fig. 10.1). The cauda equina extends from the end of the cord down to S5 within the sacral canal. Paraplegia arises from disorders affecting the thoracic spinal cord and the cauda equina, whereas quadriplegia or quadriparesis arises from disorders affecting the cervical cord. The most important information to elicit at the bedside is whether paraplegia is spastic or flaccid in type and whether there is a sensory level on the trunk above which sensation is normal (Chapter 2). This helps to localise the site of the lesion causing the paraplegia.

Key points
- paraplegia is a common neurological disorder
- clinically classified as compressive & non compressive
- main causes are Pott’s disease, transverse myelitis & malignancy
- the most common community based cause is nutritional myelopathy
- disorders of the spinal cord result in a spastic PP
- disorders of the cauda equina result in a flaccid PP
COMPRESSIVE CAUSES

The causes of compressive paraplegia can be classified on neuroimaging (CT/MRI) as being either extradural or subdural in site. Subdural includes those arising from either within the spinal cord (intramedullary) or those arising outside the cord (extramedullary) (Table 10.1). The two main compressive causes in Africa are Pott’s disease and metastatic malignancy, both of which are extradural. Acute cord compression is a medical emergency and needs rapid evaluation and intervention in order to prevent permanent disability. The history and neurological examination localise the level of weakness, and help to determine the likely cause of the paraplegia. However on their own without further laboratory and imaging investigations they may not reliably distinguish the cause.

Spinal TB
Spinal TB accounts for <10% of extrapulmonary TB. Extrapulmonary TB accounts for about 15% of all TB. Spinal TB can result in paraplegia in two main ways, either by infection of the spine (Pott’s disease) accounting for the majority of cases of paraplegia or less commonly by direct infection of the spinal cord and cauda equina (spinal cord tuberculosis).

POTT’S DISEASE
Pott’s disease is a leading cause of paraplegia in Africa and is the most common cause in childhood, adolescence and in young adults. It arises from haematogenous spread of the tubercle bacillus from pulmonary infection. The paraplegia occurs either at the time of the primary infection or more commonly 3-5 years later by reactivation. This results in infection of the spine affecting the intervertebral disc space and adjacent vertebrae which if not treated may result in characteristic extradural compression of the spinal cord causing paraplegia.

Clinical features
The main clinical features of Pott’s disease are a history of localised back pain, over weeks or months, which is made worse by weight bearing and followed by a slowly progressive paraplegia. The paraplegia may be accompanied by low grade features of active systemic TB including fever, sweating and weight loss but this is not common. Neurological examination confirms paraplegia, the clinical signs depending on the level and the extent of the lesion. The lower thoracic and the lumbar spine are the most common sites affected. The spine should be carefully inspected for signs of local tenderness and the presence of any deformity, in particular a gibbus formation. A gibbus is a visible angular deformity of the thoracic or lumbar spine caused by collapse and anterior wedging of adjacent vertebrae. Local tenderness is best elicited by gently tapping the spines of the vertebrae with a finger tip or a patellar hammer.

Investigations
The ESR is typically elevated early on and throughout the course of Pott’s disease and may be the only laboratory clue as to the likely underlying cause. Radiological evidence shows a characteristic loss of the disc space at the site of infection with destruction and wedging of adjacent vertebrae but the latter is a late finding. There may also be a paravertebral soft tissue swelling (Fig. 10.2) or uncommonly a psoas muscle abscess. A CT scan of the spine may also be helpful, particularly in early disease when plain X-rays may be normal. The diagnosis of Pott’s disease is usually made by clinical suspicion, in combination with an elevated ESR and typical...
spinal X-rays. All suspected cases of Pott's disease should have a chest X-ray to exclude active pulmonary tuberculosis.

**Treatment**
Treatment is mostly carried out on clinical and X-ray grounds alone. Where possible a needle biopsy confirming the presence of acid fast bacilli should be performed and this is diagnostic. The standard anti tuberculous treatment for spinal TB is for a total of 18 months, however a shorter 12 month course has been recently recommended. Surgical treatment involving decompression of the cord and stabilization of the spine is helpful in a few mainly early cases of spinal cord compression.

**Prognosis**
The treated mortality rate is between 10-20% and full recovery rates of from 25 to 40%.

**SPINAL CORD TUBERCULOSIS**
Direct infection of the spinal cord with TB presents clinically either as an acute myelitis developing over days or as a chronic radiculo-myelitis (Fig. 10.3) occurring over weeks to months. The resulting paraplegia is mostly lower motor neurone or flaccid in type with absent or depressed reflexes with urinary and faecal incontinence. Tuberculoma of the cord may also result in paraplegia but this is uncommon. TB meningitis may also arise from a source of infection within the spinal cord and the combination of TB meningitis and paraplegia is not uncommon in Africa. All patients presenting with paraplegia should be screened for HIV infection. The diagnosis and management of spinal cord tuberculosis is similar to that of TBM which is presented in chapter 6.
**Key points**

- Pott's disease is one of the most common causes of PP in Africa
- PP can also arise from direct TB infection of spinal cord or meninges
- Clinical features of Pott's disease are back pain, spinal tenderness & progressive PP
- Laboratory findings are a high ESR with typical spinal x-ray changes
- Treatment is with anti TB Rx for at least 12 months
- CFR is 10-20% & morbidity rate is 25-40%

**TUMOURS**

Tumours affecting the spinal cord can be either primary arising from the cord, roots or meninges or secondary or metastatic arising from outside the cord. The most common overall cause of spinal cord tumours is secondary or metastatic. The main sources are prostate, breast, lung, kidney, lymphoma and multiple myeloma. They are mostly located extradurally and the most common sites affected are the thoracic followed by the lumbar spine. These are the main cause of paraplegia in older age groups (>50 years). Primary spinal cord tumours are very uncommon with an expected incidence of <1/100,000 per year and can occur in all age groups. These include meningioma arising from the meninges, neurofibroma arising from peripheral nerves and astrocytoma and ependymoma arising from the cord.

**Clinical features**

Patients presenting with paraplegia resulting from metastatic tumours typically have a history of localised back pain in combination with a mainly flaccid type paraplegia developing over a short period, usually days or weeks. Pain arising from the spinal site of metastasis is usually...
a distinctive feature. The paraplegia may be the first presenting complaint or a complication of known systemic malignancy. In contrast benign tumours tend to present with paraplegia evolving more slowly over months or years, and pain is less pronounced.

**Investigation**
Investigations help to confirm the suspicion of a malignancy with plain spinal x-rays showing lytic or blastic lesions or vertebral collapse depending on the primary (Fig. 10.4). CSF may sometimes show a high protein level and yellow discolouration particularly if there is a block to flow resulting in a concentration of CSF constituents. This is called the Froin syndrome. Neuroimaging (CT/MRI) of the spinal cord is necessary in all patients with suspected primary cord tumours and in some with suspected metastatic tumour.

**Treatment**
The treatment depends on the type of underlying tumour. Metastatic tumours causing cord compression initially require urgent steroids and analgesia if needed. More definitive treatments include local radiotherapy and occasionally surgical decompression. Chemotherapy may be helpful in patients with lymphoma and myeloma, whereas hormonal therapy can be very beneficial in those with metastatic breast and prostate cancer. Some benign tumours can be removed depending on their histology, size and local invasiveness. The prognosis for metastatic spinal cord tumour is generally poor.

**Key points**
- metastatic malignancy is the most common cause of PP in the elderly
- prostate and breast are the most common primary sources
- paralysis is usually painful & flaccid with a sensory level on the trunk
- treatment includes steroids, analgesia, radiotherapy & chemotherapy

**ACUTE EPIDURAL ABSCESS**
Acute spinal epidural abscess is a relatively uncommon cause of paraplegia but is a medical emergency when it happens. It tends to occur in debilitated patients with diabetes, alcoholism or renal failure but may occur in an otherwise healthy person. *Staphylococcus aureus* is the most
common organism accounting for 90% of cases, skin abscesses and boils are the most common sources of infection. It can affect all age groups.

**Clinical features**
The clinical features are those of a painful sub acute paraplegia developing over hours or days. The distinguishing features are very severe pain, sometimes radicular, and local tenderness at the site of the epidural abscess which is usually situated in the cervical or lumbar spine. These are frequently but not always coupled with fever and signs of infection with or without meningism.

**Investigations**
Investigations suggest a bacterial cause when the peripheral WBC (neutrophil count) and ESR are both elevated but these can be normal. CSF examination may reveal the presence of a few white blood cells and elevated protein level or be normal. Lumbar puncture should not be performed near the suspected abscess site as this may spread the infection to the CSF causing meningitis. A CT scan of the spinal cord with contrast may show epidural enhancement but can also be normal. MRI of the spinal cord with contrast is the investigation of choice.

**Treatment**
Emergency treatment is recommended on the grounds of clinical suspicion without waiting for the results of any investigations. Appropriate high dose intravenous antibiotics including cloxacillin are started and continued for a period of up to 6 weeks. High dose intravenous steroids may be of value if given early on during the first week or two of treatment. Urgent decompressive laminectomy may sometimes be indicated.

**Key points**
- epidural abscess is a medical emergency which requires urgent treatment
- sub acute painful PP occurring over hours or days
- occurs in DM, alcoholism, renal failure & skin infection & in healthy persons
- diagnosis is confirmed by contrast enhanced CT/MRI spinal cord
- treatment is iv antibiotics & steroids & sometimes surgery

**CERVICAL SPONDYLOSIS**
This is one of the most common causes of paraplegia in high income countries but appears to be a relatively uncommon cause in Africa.

**Clinical features**
Affected patients are usually older, >60 yrs and typically present with a slow insidious onset of weakness or stiffness in the legs, sometimes coupled with numbness and tingling in the upper limbs. There may be a history of previous neck injury or accident. Pain and bladder symptoms are uncommon. The main clinical findings are those of a spastic paraplegia coupled with wasting of the small hand muscles.
Investigations
Plain X-ray or CT of the cervical spine confirms the diagnosis, showing narrowing of one or two disc spaces with prominent osteophytes, narrowing of the cervical canal and evidence of cord compression (Fig. 10.5).

Management
Management is mostly conservative and includes analgesics, cervical collar and very rarely traction. Indications for consideration for decompressive surgery include progressive paraplegia, severe weakness and intractable pain. Surgery is aimed at the prevention of any further deterioration rather than making the patient better, although neurologic improvement is occasionally seen.

Key points
- Cervical spondylosis is an uncommon cause of PP in Africa
- Presents with slow onset spastic PP with numbness & wasting in hands
- Occurs in older persons, often with a history of previous neck injury
- X-ray/CT neck: narrow disc spaces & canal with osteophytes & cord compression
- Treatment usually conservative with analgesics & cervical collar

FLUOROSIS
Fluorosis is a disorder characterized by the deposition of excess fluoride in bones (osteofluorosis) and teeth (dental fluorosis). It results from chronic ingestion of fluoride rich waters over many years. Water contamination with fluoride occurs naturally in volcanic areas of the world. This is particularly the case in volcanic areas in the rift valley region of East Africa where there can be exposure to high levels of fluoride in drinking water. The degree of water contamination and exposure can vary enormously within adjacent areas in affected regions. Neurological disorders are uncommon and arise in fluorosis as consequence of the chronic deposition of fluoride in bone occurring over many years. This can result in narrowing of the diameter
of the spinal canal and the intervertebral foramina which can cause compression. It mainly affects the cervical canal but may also affect the lumbar sacral canal. The resulting neurological disorders are mostly forms of spastic paralysis with or without associated radiculopathies. These lead to quadriplegia and paraplegia characterized by immobility, flexion deformities, severe spasms, and urinary incontinence. The diagnosis is usually suggested by the typical clinical and X-ray findings occurring in a person from a known volcanic area (Fig.10.6). Treatment is symptomatic. Primary prevention is the intervention of choice either by filtering all water at the point of access or drinking water at the point of usage.

Key points

- skeletal fluorosis results from chronic high fluoride intake in drinking water
- cervical & lumbosacral spine are most commonly affected sites
- result in a quadri/paraparesis & radiculopathy
- PP is an uncommon complication of skeletal fluorosis
- skeletal X-rays confirm the diagnosis
- prevention is by filtering drinking water

**SYRINGOMYELIA**

This is an uncommon disorder in which a CSF filled cavity called a syrinx forms in the centre of the spinal cord. The time course of symptoms and signs is one of slow progression over many years. It typically starts slowly in the lower cervical cord and over many years, the cavity can involve the whole spinal cord down to T12 & L1. The cause is considered to be related to abnormalities in the dynamics of local CSF pressure and flow. Predisposing causes include a developmental abnormality of the skull base with flattening or platybasia coupled with variable protrusion of the lower brain stem and cerebellar tonsils through the foramen magnum. This is called the Arnold-Chiari malformation. Neck and cervical cord injury is another risk factor which is an important cause in Africa, particularly in women because of falls whilst carrying...
heavy loads on the head. There is usually a gap of 20-30 years from the time of the accident to the onset of first symptoms.

**Clinical features**
Syringomyelia results in a characteristic pattern of motor and sensory deficits with signs of a spastic paraplegia in the legs, and lower motor neurone signs in the upper limbs. Pain is a feature; this is often severe, burning or neuropathic in type and often in a radicular distribution in the limbs. The sensory findings are diagnostic, with involvement of the spinothalamic tracts producing loss of pain and temperature in a characteristic cape distribution over the shoulders. Sensory involvement occurs in the arms and legs depending on the site and extent of the syrinx. Because the cavity is situated in the centre of the cord there is sparing of the posterior columns with preservation of joint position, vibration and light touch. Characteristic clinical findings include neuropathic scars because of the loss of pain sensation, wasting of small hand muscles and claw hand deformities (Fig. 10.7). The clinical course in severe cases is a slowly progressive quadriplegia or paraplegia occurring over many years with the patient gradually becoming wheelchair and bedbound.

**Treatment**
Treatment is usually conservative. Neurosurgery, if indicated is aimed at stabilizing the condition with decompression of the foramen magnum and drainage of the syrinx.

![Neuropathic scars and clawing](Figure 10.7 Hands in syringomyelia)

**Key points**
- Syringomyelia is uncommon cause of PP caused by a cavity within the spinal cord
- Previous spinal injury & platybasia are the main risk factors
- History of injury is often >20 years earlier
- CFs are burning pains & wasting & neuropathic scars on the hands/arms
- A slowly progressive SPP with loss of pain & temp but joint position/vibration are intact

**NON COMPRESSIVE**
The main non compressive causes of paraplegia in Africa are transverse myelitis, HIV, TB, schistosomiasis, syphilis, B-12 deficiency and HTLV-1.
TRANSVERSE MYELITIS (TM)

Transverse myelitis is a term used to describe an episode of inflammation affecting the spinal cord which results in an acute paraplegia. It is one of the major causes of paraplegia in young and middle aged persons in Africa. Mechanisms are considered to be infectious and autoimmune in origin, but in most cases the exact cause is not known. Viruses known to be involved are herpes zoster, herpes simplex, HIV and HTLV-1. Devic’s disease (neuromyelitis optica) and systemic lupus erythematosis (vasculitis) are uncommon causes.

Clinical features

Transverse myelitis typically results in an acute devastating flaccid paraplegia, developing over hours or days with a sensory level on the trunk and loss of control of bladder and bowel. Pain though frequently present at onset is mild and usually clears. Spasticity usually develops later, but may not in very severely affected cases who remain flaccid because of involvement of the anterior horns in the spinal cord. At onset the CSF may show elevation in leucocytes and protein and MRI imaging may show high signal in the cord (Fig.10.9). CT examination of the spine is useful to exclude other causes.

Treatment

Treatment initially is with high dose intravenous steroids in combination with a course of antiviral medication e.g. acyclovir 10 mg/kg (800 mg) iv/po/3-4 times daily for 10 days if clinically appropriate. High dose steroids include methylprednisolone 1000 mg iv daily for 5 days followed by oral prednisolone 60 mg daily, tapering over 2-3 weeks. High dose dexamethasone, 24-32 mg daily can also be used if methylprednisolone is unavailable.

Prognosis

Recovery is variable, it may be rapid but in some cases can be slow and incomplete, occurring over months or years. However, most patients are permanently disabled and confined to a wheelchair.

Key points

- transverse myelitis is one of the common forms of PP in Africa
- affects mainly young & middle aged persons
- viruses are a known cause but in most cases the cause is not identified
- paraplegia occurs over hours or days with initial flaccidity, sensory level & incontinence
- treatment: antiviral medication & iv steroids but most persons are left permanently disabled

HIV

Paraplegia is a major neurological disorder in HIV disease (Chapter 8). It can occur during the asymptomatic stage of HIV infection when CD4 counts are >200/cm³ and more commonly during the symptomatic stage when CD4 counts are very low <100/cm³. The main causes are opportunistic processes and direct HIV infection of the spinal cord. Opportunistic infections (OIIs) include tuberculosis, herpes zoster, herpes simplex, CMV, syphilis and co-infection with HTLV-1 in endemic areas. The main opportunistic tumour is lymphoma.

Direct infection of the spinal cord causes a vacuolar myelopathy (VM) in advanced HIV infection. (Chapter 8). This results in a progressive spastic paraplegia associated with urinary
incontinence and sensory ataxia in <1% of patients. More common findings in vacuolar myelopathy are isolated hyperreflexia in the legs, usually the knees and extensor plantar reflexes. These isolated neurological signs are present in >20% of patients with advanced disease. Infection with cytomegalovirus can uncommonly cause a paraplegia during the later stages of HIV infection. This is due to lumbosacral radiculopathy and results in a flaccid paraplegia. Management of paraplegia in HIV is directed at finding and treating the underlying cause and starting ART depending on the degree of immunosuppression.

Key points
- PP is a major but a relatively uncommon neurological complication in HIV disease
- main mechanisms are OIs & direct HIV infection of the cord
- main OIs are TB, viral infections & syphilis
- vacuolar myelopathy arises from HIV infection of cord & causes PP in <1% of pts
- management is by treating the underlying cause & by starting ART

SCHISTOSOMIASIS
Involvement of the spinal cord is considered to be uncommon, although 1-5% of all cases of non traumatic paraplegia in schistosome endemic parts of Africa are reported to be caused by schistosomiasis (Chapter 7). Reports from schistosome affected areas of Malawi suggest that it may be even more common there. Paraplegia occurs mostly with *S. mansoni* (SM) and occasionally with *S. haematobium*. It happens particularly in early infection in the non immune host and when there is a heavy worm and egg load.

Pathogenesis
The paraplegia arises because of marked acute inflammatory response of the host to schistosome eggs being deposited in and around the spinal cord (Fig. 10.8). The likely source of the eggs are ectopic adult worms either living in veins around the cord or by retrograde flow of eggs through valveless spinal veins in connection with the iliac veins. The time between exposure and onset of the paraplegia is usually weeks to months and rarely years. The conus medullaris and the cauda equina are the most common sites affected (Chapter 7).

Clinical features
The clinical features include lumbar pain followed by difficulty passing urine, which may precede the other symptoms by days or weeks. The main presentation is that of a progressive flaccid paraplegia, occurring over days or weeks, usually with sensory and bladder involvement. Other neurological presentations include myeloradiculopathy and spastic paraplegia.

Diagnosis
The diagnosis should be considered in any patient with an acute onset paraplegia who is living in or has come from an endemic area. The diagnosis is difficult because the paraplegia mainly occurs during the early invasive phase of the adult worms, when there is little clinical or laboratory evidence of underlying schistosome infection. Stool examination for eggs may be negative and rectal snips are positive in only about 50% of cases. A positive serological test is useful in non endemic patients as an indicator of previous exposure. CSF examination may show lymphocytes and occasionally eosinophils as well as an elevation in protein. CT of the
spinal cord is usually not helpful but MRI imaging with contrast may show hyper intense areas in the cord or irregular enhancement and dilation of the conus.

**Figure 10.8** Schistosome mansoni eggs in the spinal cord, histopathology

### Treatment

Treatment is with praziquantel 40 mg/kg/day orally for up to 14 days in combination with oral prednisolone 60-90 mg daily for 3 weeks. Surgery has been tried for acute cases of failed medical treatment. The prognosis is guarded, with about 10% mortality, 30% remaining permanently paraplegic and 60% showing moderate to good recovery.

### Key points

- Schistosomiasis is a known cause of acute PP in endemic areas
- Lower end of spinal cord & the cauda equina are the main sites affected
- Flaccid PP is the main clinical presentation
- Diagnosis is difficult because there is little laboratory evidence of infection
- Treatment is with praziquantel & steroids for 2-3 weeks
- CFR is 10% & 30% left paralysed

Other causes of non compressive paraplegia include syphilis, human T cell lymphotropic virus type 1 (HTLV-1), Devic’s disease, vitamin B-12 deficiency, ischaemia, sickle cell disease, vasculitis, arteriovenous malformation (AVM) and hereditary spastic paraplegia.

### Syphilis

Syphilis is a sexually transmitted disease caused by the spirochete *Treponema pallidum* (Chapter 6). It is estimated that neurological involvement will occur in about 7% of patients 10-25 years after primary infection, if primary syphilis is untreated. However despite the frequency of primary syphilis in Africa, neurosyphilis remains uncommon in many countries there. The reason for this has been ascribed to the continued widespread use of antibiotics inadvertently treating early infection or altering the natural history of clinical disease. The incidence of neurosyphilis is increased when syphilis is associated with HIV infection.
Clinical features
Spinal syphilis typically presents either as a progressive spastic paraplegia or as tabes dorsalis with posterior column loss, lightning pains and Charcot’s joints. However tabes dorsalis remains a distinctly uncommon clinical presentation in Africa.

Diagnosis
The diagnosis of neurosyphilis depends on the serological detection of antibodies in both blood and CSF. The Venereal Disease Research Laboratory (VDRL) is the screening test most commonly used in Africa although it may be negative in HIV infection. Limitations to VDRL include false positives in serum but not in the CSF which can however be negative in 20-30% cases. More sensitive and specific diagnostic antibody tests include the fluorescent treponemal antibody absorption (FTA) and the treponemal antibody immobilization test (TPI). The CSF may also show elevated leucocytes and protein.

Treatment
Treatment is with penicillin 2-4 million units i.v. 4 hourly for 14-21 days.

HUMAN T CELL LYMPHOTROPIC VIRUS TYPE 1 (HTLV-1)
HTLV-1 is a retrovirus which is endemic in some areas of western, southern and central Africa with just a few clusters reported in eastern Africa. It is endemic in areas of Japan, the Caribbean and South America. The serological prevalence rates are 3-6% in some of the worst affected communities. It is transmitted perinatally, sexually and by blood transfusion. Chronic infection for up to 20-30 years can result in a slow progressive form of tropical spastic paraplegia known as HTLV-1 associated myelopathy (HAM). However this occurs in only 1-5% of persons infected with the virus with the majority being asymptomatic carriers. The distinctive clinical feature of HTLV-1 is that of a progressive spastic paraplegia characterized by slow onset over years of stiffness and weakness affecting the legs with paraesthesiae, back pain and urinary symptoms. The diagnosis is confirmed by finding HTLV-1 antibodies in the blood and CSF. There is no effective treatment although steroids have been tried.

DEVIC’S DISEASE (NEUROMYELITIS OPTICA)
This is an uncommon but severe disease in Africa characterized by recurring attacks of acute severe demyelinating transverse myelitis and optic neuritis. This results in paraplegia and blindness. Occasionally, the disease is monophasic but this appears to be unusual in Africa. It occurs predominantly in females in their late teens or early 20s in Africa but may affect older women in their 30s and 40s in high income countries. The optic neuritis typically precedes the transverse myelitis but can also occur concurrently with it. The relapses are severe, occurring usually within 6 months of the first and subsequent episodes.

Diagnosis
The diagnosis in Africa is mainly clinical. Examination of CSF may show increased protein and the presence of lymphocytes. A highly specific immunoglobulin antibody to aquaporin (NMO –IgG) can now be detected in blood and CSF of infected persons but the test is only available in some specialised neurological centres in Africa. MRI imaging if available shows increased signal in the spinal cord often spanning several vertebral levels and almost complete withering of the cord in cases of long standing disease (Fig. 10.9).
Treatment
Treatment for the acute attacks is with high dose steroids for periods of 4-6 weeks and if available, repeated plasma exchange or intravenous immunoglobulin for the non responders. Long term prophylaxis with a combination of steroids and azathioprine or other forms of immunosuppression has been tried but the response is variable. Residual disability after an attack is usual with many patients becoming wheelchair or bed bound within 1 to 2 years of onset.

VITAMIN B-12 DEFICIENCY
Vitamin B-12 deficiency causes a peripheral neuropathy, sub acute combined degeneration of the cord (SACD), optic atrophy, dementia and pernicious anaemia. The main spinal findings are those of a myelopathy with brisk knee reflexes and up going plantars. These typically occur in combination with signs of neuropathy, including absent ankle jerks and loss of peripheral sensation including joint position sense. All or some of the above may be present in any one patient. The main causes of SACD are nutritional deficiency, seen most commonly in vegetarians, and malabsorption. Malabsorption may be caused either by a lack of intrinsic factor which seems to be quite uncommon in Africa and also by diseases of the terminal ileum.

Diagnosis
Diagnosis is by finding the typical neurological findings in combination with evidence of megaloblastic anaemia i.e. elevated mean corpuscular volume (MCV) and low serum B-12.

Treatment
Treatment is with intramuscular injections of B-12. The dose is 1 mg on alternate days for a total of five injections followed by 1 mg injections every 3 months for life. If intramuscular injections are not available then B-12 can sometimes be given orally, in a dose of 1 mg po daily.
This may provide adequate replacement particularly in cases secondary to nutritional or dietary deficiency.

**Key points**

- neurosyphilis is uncommon in many countries in Africa
- HTLV-1 is endemic in selected areas of South, West & Central Africa
- Devic’s disease is uncommon but severe demyelinating disorder of CNS
- B-12 deficiency is a treatable but uncommon cause of neuropathy & paraplegia

**TROPICAL MYELONEUROPATHIES**

These are a group of paraplegias and peripheral neuropathies that are considered to be nutritional in origin. They include the tropical spastic paraplegia (TSP) group, konzo, and lathyrism, and the peripheral neuropathy group, tropical ataxic neuropathies (TAN). These disorders represent a distinct group of community based myeloneuropathies which result in paraplegia. They occur largely as epidemics and are the commonest cause of neurological disability in the rural communities in which they occur in Africa. These are non curable when they happen and their management lies mainly in prevention.

**KONZO**

Konzo is a distinct form of tropical myeloneuropathy characterized by abrupt onset of a non progressive but permanent spastic paraplegia related to cassava consumption. It occurs mainly as epidemics in exclusively cassava growing areas of the east, central and western Africa. Epidemics have been known to occur in cassava growing of the former Belgian Congo as far back as 1928 and possibly earlier. In epidemics as many as 1-30/1000 persons are affected, mainly growing children and fertile women. It also occurs in an endemic form but at much lower rates.

**Aetiology**

The cause of konzo has been attributed to the combined effect of months of high cyanide and low protein (methionine and cysteine, sulphur based amino acids) intake from exclusive consumption of insufficiently processed bitter cassava (Fig. 10.11). The bitter cassava grow well in poor soils but contain increasing amounts of cyanogenic glycosides mainly linamarin. Processing disrupts the root tissue and releases volatile hydrogen cyanide and this makes the food safe for human consumption. The main processing methods used in Africa involve hydrolysis or soaking in water, crushing and fermentation or sun drying. Safe processing takes days in the case of hydrolysis and fermentation, to weeks for sun drying. Shortening of these methods is a risk factor for the disease.

**Clinical features**

The clinical features are characterized by an abrupt (usually <24 hours) onset of a permanent but non progressive spastic paraplegia (Figs. 10.10). The upper limbs and optic nerves may also be involved in some cases. The bladder, bowel and sensation are all spared. The range of disability varies from very mild upper motor neurone findings in the legs to spastic paralysis of all four limbs in very severe cases. The majority of persons with konzo can walk with the aid of crutches.
Diagnosis
The clinical criteria for diagnosis are an abrupt (<1/52) onset of spastic paraparesis in the absence of any other cause in a patient coming from a cassava growing area. Supportive investigations include elevated blood or urine thiocyanate levels and low levels of the essential amino acids methionine and cysteine. These investigations are only available in specialist or research laboratories.

Clinical criteria for diagnosing konzo
- spastic paraplegia
- abrupt onset in <one week
- occurring in a cassava growing area
- no other cause is found

Treatment and prevention
There is no medical treatment for the condition. Prevention is mainly directed at growing cassava with lower cyanide content and public education concerning safer methods of cassava processing. A newer and safer “wetting method” of cassava processing has recently
been promoted in some affected communities in East Africa. Additional measures include establishing early warning systems of potentially high cyanide levels in cassava tubers and of pending epidemics and the provision of supplementary protein before or early on during epidemics. Surgical treatment involving Achilles tendon lengthening operations have proved successful in increasing mobility in some patients.

**LATHYRISM**

Lathyism is an epidemic form of spastic paraplegia found almost exclusively in parts of India, Bangladesh and South East Asia where large quantities of the grass pea (*Lathyrus sativus*) a drought resistant crop are grown and consumed. In Africa it is found only in parts of North Western Ethiopia where the grass pea is widely grown and consumed. The association of paraplegia with grass pea consumption was already recognized in ancient Greece.

**Aetiology**

The disease is caused by a neurotoxic amino acid found in the grass pea called beta-N-oxalylamino-L-alanine (BOAA). Lathyism occurs after weeks or months of almost exclusive consumption of the grass pea.

**Clinical features**

It occurs mostly as epidemics and occurring with about the same frequency and age distribution as Konzo. It is clinically almost identical to konzo, apart from the mild sphincter involvement occasionally found in lathyism (Fig. 10.12). However, they are distinguishable from each other because they both occur in geographically distinct areas which do not overlap.

**Treatment and prevention**

The management and prevention are along the same principles as that for konzo.
Key points

- konzo and lathyrism are distinct forms of tropical spastic paraplegia
- occur mainly as epidemics in different areas in Africa
- konzo occurs only in cassava growing & lathyrism occurs only in grass pea growing areas
- each is caused by weeks/months of almost exclusive consumption of a staple food
- both result in permanent spastic paraparesis & are prevented by similar measures

TROPICAL ATAXIC NEUROPATHY (TAN)

This is another distinct form of tropical myeloneuropathy. It has originally been described in cassava eating populations in Nigeria and Tanzania in the 1960s and more recently again in Nigeria and in southern India. The disease is characterized by a combination of gradual onset in older adults of peripheral neuropathy, sensory ataxia, optic neuritis, deafness and sometimes spastic paraplegia. The presence of any two or more of these findings is sufficient to make the diagnosis. The cause is not known but is linked to chronic cyanide exposure from chronic cassava consumption in combination with possible vitamin B nutritional deficiency. Management includes vitamin B supplementation.

MANAGEMENT OF PARAPLEGIA

There are four major principles governing the management of patients presenting with paraplegia.

**Do not delay**

The early establishment of correct diagnosis, treatment and the prevention of complications are critical to the outcome for the patient. It is important to preserve whatever neurological function is still remaining.

**Establish the cause**

The cause is established by history, physical examination, laboratory and radiological investigations. The main investigations are outlined below in Table 10.2.

Table 10.2 Main investigations in paraplegia

<table>
<thead>
<tr>
<th>Type of investigation</th>
<th>Investigation</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Bloods</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Haematology</strong></td>
<td>FBC, ESR, B-12, sickle cell test</td>
<td>TB, malignancy, SACD, sickle cell disease, abscess</td>
</tr>
<tr>
<td><strong>Biochemistry</strong></td>
<td>glucose, creatinine, electrolytes, calcium, total protein and electrophoresis</td>
<td>myeloma, malignancy</td>
</tr>
<tr>
<td><strong>Serology</strong></td>
<td>HIV, VDRL, schistosome, cysticercosis and echinococcosis serology</td>
<td>HIV disease, syphilis, helminthic infections</td>
</tr>
</tbody>
</table>

*continues*
Treat the cause
The treatments for the main causes of paraplegia are outlined below in Table 10.3. There is a low threshold for starting TB treatment in Africa, particularly in younger patients presenting with paraplegia.

<table>
<thead>
<tr>
<th>Disease</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Potts disease</td>
<td>TB Rx</td>
</tr>
<tr>
<td>TB spinal cord</td>
<td>TB Rx &amp; steroids</td>
</tr>
<tr>
<td>metastatic malignancy</td>
<td>steroids &amp; analgesia</td>
</tr>
<tr>
<td>transverse myelitis</td>
<td>steroids &amp; acyclovir</td>
</tr>
<tr>
<td>HIV</td>
<td>treat the cause &amp; ART</td>
</tr>
<tr>
<td>cervical spondylosis/disc</td>
<td>surgical decompression if indicated</td>
</tr>
<tr>
<td>schistosomiasis</td>
<td>praziquantel &amp; steroids for 2 weeks</td>
</tr>
<tr>
<td>syphilis</td>
<td>penicillin for 21 days</td>
</tr>
<tr>
<td>Devic's disease</td>
<td>steroids</td>
</tr>
<tr>
<td>SACD</td>
<td>B-12</td>
</tr>
<tr>
<td>acute epidural abscess</td>
<td>iv antibiotics</td>
</tr>
</tbody>
</table>

Prevent complications
The patient needs assistance to care for skin, limbs, bladder and bowels. This is best achieved by the early involvement of the patient’s family in cooperation with the nurses and physiotherapists.

Skin
The prevention of pressure sores is critical to the patient’s survival. In immobile patients pressure sores develop quickly over bony prominences particularly the sacrum, hips and heels. This is more likely to occur during the first days of the illness. Specially designed air mattresses that minimize this risk are unavailable in most hospitals in Africa. Prevention of bed sores can best be achieved by a rigorous approach which involves 2 hourly turning of the patient day and night and starting immediately on admission. This task should be entrusted to a family carer or carried out by the nursing staff.

The skin should be inspected frequently and always be kept dry and clean. Attention early to adequate nutrition and measures to prevent infection are also very important. These measures include the use of mosquito bed nets to prevent malaria and a bed cage to keep weight of blankets off the paralysed legs. If bed sores do become established they require vigorous cleaning, surgical debridement and possible skin grafting.
**Bladder and bowels**

Urinary catheterization is necessary when there is a non-functioning bladder. This typically occurs during the first days of an acute paraplegia and persists particularly in patients with flaccid paraplegia. The bladder is innervated by both the autonomic and the somatic or voluntary nervous system. Loss of control of bladder function or neurogenic bladder arises because of lesions situated in either the spinal cord or cauda equina. Patients with spinal cord lesions and usually a spastic paraplegia may eventually after months develop satisfactory reflex bladder emptying or require intermittent self catheterization and treatment with antispasmodic or anticholinergic drugs. Patients with a cauda equina lesion and flaccid paraplegia usually require permanent catheters. It may be necessary to use antibiotics for urinary tract infections.

Constipation is a feature of both flaccid and spastic paraplegia with the bowels opening every week or less often. Measures to prevent it include satisfactory fluid intake, adequate fibre/bulk in the diet, and the early use of laxatives usually the combination of a lubricant and an irritant. Manual evacuation may be necessary when there is faecal impaction.

**Limbs**

Care of paralysed limbs involves frequent passive movements. This helps to prevent thrombosis, joint stiffness, spasticity and contractures, and to exercise the non-affected muscles. The management of spasticity and avoidance of contractures is best done by a physiotherapist and trained family members, and also through the use of antispasmodics.

The main antispasmodics available are baclofen and diazepam. The starting dose of baclofen is 5 mg twice daily increasing slowly over weeks to 20-40 mg twice daily as required. The limiting adverse effects are drowsiness, fatigue, and the fact that it can sometimes take away the spasticity that the patient needs and uses to be able to stand and mobilise. The starting dose of diazepam is 2-5 mg three times daily, increasing to a maximum of 20 mg three times daily. The limiting adverse effects are drowsiness and fatigue, and anxiety may occur on sudden withdrawal. Other drugs used for spasticity include dantrolene and tizanidine. Further measures include the use of support stockings and low dose heparin which help to prevent deep vein thrombosis.

**Key points**

- treat the cause without delay & aim to prevent complications
- main complications are bedsores, urinary retention, infection & spasticity
- 2 hourly turning should start immediately on admission
- entrust care to a relative or carer

**LONG TERM MANAGEMENT**

Paraplegia is a devastating illness which results in disappointment, depression, shame, resentment, anger and an altered role in family. Patients with chronic paraplegia have to come to terms with the loss of function of the lower half of their bodies. They need support in the activities of daily living (ADL), mobility, work and family life. They benefit particularly from specialized knowledge, support and encouragement. In a medical setting, this is usually provided by medical staff including nurses, doctors, physiotherapists, occupational therapists and carers. In order to cope at home, the person and their family need the wider support of carers, friends and the community. Longer term measures should include plans for social and...
recreational activities and employment in order for the patient to be more able to participate as fully as possible in everyday life. Some specific measures are outlined below.

**Motor**
The provision of a wheelchair is often both necessary and essential for mobility. The patient needs to be educated about the use of a wheelchair and learn the skills needed for independent transfers. This will involve the patient learning some knowledge about the level of cord involvement and injury and the extent and severity of any resulting paralysis and disability. The patient needs to train to strengthen the non paralysed muscles and family members need to be instructed and trained to regularly passively move paralysed limbs in order to prevent contractures and bed sores. House adaptation is usually necessary for use of a wheelchair.

**Skin**
Great care needs to be taken to prevent the development of pressure sores. A routine of avoiding prolonged periods of weight bearing will need to be established and maintained. This is helped by the person regularly taking the weight of the body off the seat of chair or wheelchair a couple of times every hour and by using protective cushions to guard against pressure points. Regular inspection of skin is essential and may of necessity have to be carried out by the carer.

**Bladder and bowels**
The patient needs training in reflex bladder emptying, condom drainage, intermittent self catheterization, indwelling catheter management, anticholinergic drugs and early recognition and treatment of urinary tract infections. The patient needs a healthy wholesome and regular diet with laxatives and suppositories to avoid constipation.

**Sex and fertility**
This needs to be discussed and explained with the patient and partner. Erectile potency may be retained in upper motor neurone spinal cord disorders and respond to oral medications phosphodiesterase inhibitors e.g. sildenafil. In contrast erectile potency is lost in cauda equina or lower motor neurone disorders. In female patients local spasticity in the thigh adductors may be helped by local antispasmodic injections or medications.

**Weight and calories**
Paraplegic patients need on average 40-50% less calories. This needs to be explained clearly to the patient in order to avoid excessive weight gain.

**Key points**
- assist & support the person to recover functional independence
- involves prevention of bed sores, urinary tract infection & weight gain
- includes provision, use and maintenance of a wheelchair
- long term aim is to participate as fully as possible in everyday life
- needs support of carers, family, friends & community
Selected references


