

PARALYSIS - CHRONIC - FLACCID

Flaccid (hypotonic) paralysis of gradual onset is usually due to peripheral nerve disease, but it may be caused by a lesion of the anterior horn cells, motor roots, peripheral nerves, motor end plates or muscle. The reflexes are usually preserved until late in the course of muscular and the neuro-muscular junction diseases.

CEREBRAL LESIONS

Focal cerebral lesions usually cause unilateral weakness with increased tone, increased reflexes, upgoing plantars and no sensory loss. However, there may be initial hypotonia with an acute cerebral lesion.

Generalised chronic cerebral lesions may cause hypotonic weakness (which is unexpected, because upper motor neurone lesions usually cause hypertonia), but there are brisk reflexes (as expected in upper motor neurone lesions):

- encephalopathy (damage to normally formed brain): atonic cerebral palsy (atonic diplegia) is the commonest cause of the “floppy infant syndrome”; the plantars are usually upgoing
- dysgenesis (abnormally formed brain): eg Down’s syndrome, Prader-Willi syndrome
- degenerative (regression of normal CNS): storage diseases of lipid, mucopolysaccharide etc. There is a period of normal development, followed by loss of milestones.

SPINAL CORD LESIONS

Occult spina bifida may cause flaccid paraplegia. Spinal cord compression in the lumbar region may cause flaccid paralysis due to a cauda equina lesion. Acute compression of the thoracic or cervical cord may cause flaccid paralysis for a time (spinal shock), but compression of gradual onset causes spastic paralysis.

ANTERIOR HORN CELL DISEASE

Werdnig-Hoffmann disease is a rare progressive form of motor neurone disease that causes progressive hypotonia with absent reflexes and fasciculations. It usually starts in infancy.

PERIPHERAL NERVE LESIONS

Distal weakness with early impairment of reflexes, hypotonia, paraesthesiae and sensory loss. The diagnosis may be confused by the initial stages of a spinal lesion (spinal shock) or acute hemiplegia before hyperreflexia occurs, and by advanced muscle disease. If a child has no reflexes despite reinforcement (biting hard, pulling hands apart), he usually has neuropathy.

There are many causes of neuropathy. These include toxic substances (tick bite, heavy metals), drugs (such as isoniazid, streptomycin, phenytoin, chloroquine, vincristine, DDT), deficiency diseases (beriberi, malabsorption of B12 and folic acid), infections (leprosy, diphtheria, tetanus, dysentery, malaria, TB, typhoid), post-infective (Guillain-Barre), trauma, systemic disease (SLE, PAN, rheumatoid, sarcoid), genetic (peroneal muscular atrophy, Refsum’s disease) and idiopathic.

MYONEURAL JUNCTION DISORDERS

The classic cause is myasthenia gravis. Symptoms include ptosis, ophthalmoplegia, generalised weakness, dysphagia and occasionally respiratory paralysis. The reflexes are normal. There is a transitory neonatal form in the babies of affected mothers. Aminoglycoside antibiotics and hypermagnesaemia (seen in the babies of mothers given magnesium for eclampsia) may affect the myoneural junction.

MUSCLE DISEASE

There is usually gradual onset of proximal muscle weakness. The child “climbs up” his legs with his arms when going from lying to standing because of thigh weakness (Gower’s sign). Neck flexion is weaker than neck extension. Reflexes are normal until late in the disease. Sensation is normal. There may be wasting, but there are no fasciculations. The CSF is normal.

Check for facial involvement (facioscapulohumeral dystrophy), rapid onset of fatigue (myasthenia), pelvic and thigh muscles worse than head and shoulders (limb-girdle dystrophy), myotonia, hypertrophy (Duchenne dystrophy).

Serum CPK, CSF examination and muscle biopsy may help in arriving at a correct diagnosis.

BENIGN CONGENITAL HYPOTONIA

This is a diagnosis of exclusion.

PARALYSIS - SPASTIC (HYPERTONIC)

Spastic paralysis implies disease of the brain or spinal cord (upper motor neurone lesion), with hyperreflexia and upgoing plantars. Active reflexes in a child with downgoing plantars are usually normal. Upgoing plantars are normal in children up to 12 months old.

A child with recent onset of focal spastic paralysis (suggesting a cerebral space occupying lesion) should have a trial of TB therapy, unless neoplasm has been proven histologically.

- Unilateral:
- previous injury - birth, meningitis, subdural haematoma, trauma
 - recent onset - investigation needed to exclude cord compression (Brown-Sequard) or cerebral tumour, tuberculoma, toruloma, haematoma or subdural effusion.
- Bilateral:
- cerebral palsy (spastic diplegia)
 - spinal cord compression (local back pain, sensory level) - familial spastic paraplegia (?family history)
 - spastic tropical paraplegia
 - subacute sclerosing panencephalopathy (with ataxia) (p.358)
- In arms and legs:
- lesion in cervical cord or higher
- In legs only:
- the lesion is below the cervical cord EXCEPT FOR:
 - cerebral palsy (leg fibres selectively damaged by hypoxia) giving spastic diplegia
 - hydrocephalus (parasagittal leg fibres stretched most by dilated ventricles)
 - parasagittal intracranial mass, eg meningioma (presses on area of cortex controlling legs); look for headache, personality change, convulsions.

CEREBRAL SPACE-OCCUPYING LESION

A lesion of the brain usually causes hyperreflexic spastic paralysis (with an upgoing plantar) on the opposite side of the body (though an acute lesion may cause flaccid paralysis initially). There may be raised intracranial pressure (see p.77). Lesions of the cerebral cortex and internal capsule cause paralysis of the face, arm, and/or leg (depending on the extent of the lesion) on the opposite side of the body, but only very large lesions in the anterior fossa cause raised intracranial pressure (with small ventricles). Lesions in the posterior fossa may cause weakness of the face on the same side as the lesion (or on the opposite side) and weakness of the arm and leg on the opposite side, and quite small lesions may cause raised intracranial pressure due to obstruction of the flow of CSF (with enlarged ventricles). Neck stiffness in a patient with focal neurological signs of gradual onset suggests a posterior fossa lesion, or meningitis due to TB or cryptococcus. Subarachnoid haemorrhage and purulent meningitis cause neck stiffness of acute onset.

Do NOT do a lumbar puncture if there is any evidence of raised intracranial pressure.
Do a chest x-ray and Mantoux.

The most likely space-occupying lesion in Papua New Guinea children is a tuberculoma. It is therefore always justified to give a trial of antituberculous treatment.

If there is no improvement after four weeks of anti-TB therapy, or if there is deterioration prior to this, a search for other lesions may be considered - although it should be remembered that the prognosis for intracranial neoplasms in children is poor. Before embarking on invasive and expensive investigations, it is important to discuss the implications with the family.

The availability of a CT scan facility in Port Moresby has made the diagnosis of intracranial neoplasms in all age groups possible by non-invasive means. In young children with "sprung" sutures it may be possible to perform ultrasound scan of the brain through the sutures.

Should these investigations be impracticable, invasive investigation such as burrhole and needle exploration, and air ventriculogram might be considered.

Administration of steroids is likely to produce temporary improvement in symptoms and signs related to intracranial neoplasm, as a result of reduction of inflammatory oedema.

In situations where the prognosis is bad, parents may well wish to take their child home to die rather than stay in hospital.

Pain palliation is something which doctors can help with - even in "hopeless" situations.

TUBERCULOUS SPINAL OSTEITIS (POTT'S DISEASE)

Affected children may present with local pain and tenderness of the spine (usually in the thoracic region), or they may present with difficulty in walking.

NEVER IGNORE BACK PAIN OR LOWER LIMB WEAKNESS OR LIMP IN CHILDREN.

There is often hyperreflexia and sensory loss in the legs, a visible deformity of the spine (gibbus) and a positive Mantoux. Chest x-ray may be suggestive of tuberculosis, and x-ray of the spine almost always shows erosion and collapse of one or more adjacent vertebral bodies. Treatment is with antituberculous drugs.

The place of surgery in spinal cord compression secondary to Potts disease has been controversial, but current opinion is that it is indicated only if the signs have been rapidly progressive and the disability is severe.

SPINAL ADHESIVE ARACHNOIDITIS

This is a disease of older children and adults (usually male). Patients usually give a history of backache followed by weakness and sensory loss which develops over some weeks. In most cases, the lesion is in the thoracic or cervical spine, and there is spastic paraplegia. In some patients, the lesion is in the lumbar spine, and there is flaccid paralysis due to a cauda equina lesion. Myelogram usually shows an irregular block and the diagnosis is confirmed at laminectomy. The prognosis for recovery is very poor. Steroids should not be given (see Wagner F. PNG Med J 22:57-61,1977. Spinal adhesive arachnoiditis).

SPASTIC TROPICAL PARAPLEGIA

This is a progressive myelopathy probably caused by the HTLV-1 virus. HTLV-1 virus is widespread in Papua New Guinea.

PERICARDIAL EFFUSION AND PERICARDIOCENTESIS

The widespread availability and use of ultrasound has confirmed clinical impressions that pericardial effusions are relatively common. Many are small, contribute little to the clinical presentation and resolve on treatment of the underlying condition with no sequelae. Some, however, are large and cause compression of the heart with impairment of function - TAMPONADE. Some are purulent and require drainage of pus. Others, if not drained, may, in the long term, result in fibrosis and constrictive pericarditis.

AETIOLOGY

Causes of pericarditis include:

- viral infection
- staphylococcus*
- uraemia
- SLE
- tuberculosis
- rheumatic fever
- rheumatoid arthritis
- leukaemia

* spread from pneumonia or septicaemia

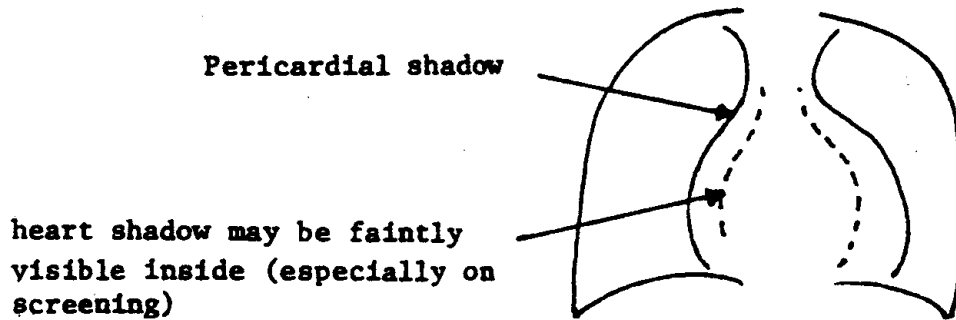
DIAGNOSIS

Ultrasound should be performed whenever there is a suspicion of pericardial effusion. It is vital to diagnose tamponade and purulent pericardial effusion and to drain the pericardium. Tamponade may be mistaken for myocarditis or heart failure due to pneumonia. Tamponade is suggested by Beck's triad (quiet heart, rising JVP, falling BP) in an anxious, ill child. However, many children with tamponade do not have muffled heart sounds.

In cardiac tamponade:

1. there is a rapid pulse, usually with pulsus paradoxus - a small volume pulse that is even weaker on inspiration. The systolic blood pressure usually falls during inspiration, but a fall of over 10 mmHg is abnormal (but not really "paradoxical" at all). It occurs in tamponade, airway obstruction (such as in severe asthma) and SVC obstruction
2. the JVP is ALWAYS raised, though it may be so high that it is hard to see. It may rise further on inspiration
3. the apical impulse is usually markedly reduced (unlike cardiac failure), but this may not be so
4. the heart sounds may be quiet. The presence of a third heart sound or pericardial rub suggest cardiac failure rather than tamponade (but may be present in tamponade)
5. tender hepatomegaly may develop, but is absent initially. Peripheral oedema is rare
6. ECG often shows reduced voltages with flattened T waves
7. chest x-ray usually shows cardiomegaly, but this may not be gross in acute tamponade, which can be caused by quite a small effusion. Pulmonary oedema is very rare
8. ultrasound screening shows a collection of fluid (dark area) around the heart. If it is a serous effusion, it will be black. If it is pyogenic, it may have a speckled greyish appearance and if it is a tuberculous fibrous exudate, there may be readily visible fibrous strands and clots. There is likely to be reduced movement of the heart, though this is more difficult to appreciate for the non expert.

Chest x-ray with pericardial effusion



ALL PRACTICING CLINICIANS SHOULD BE ABLE TO DIAGNOSE A PERICARDIAL EFFUSION USING ULTRASOUND.

Other investigations may be indicated:

- FBE (elevated WCC may suggest pyogenic effusion in a toxic child)
- blood culture
- serum urea
- LE cells and ANA
- Mantoux.

TREATMENT

Do NOT give digoxin or diuretics.

PERICARDIOCENTESIS is indicated if:

- there is tamponade
- there is the possibility of pus in the pericardium.

The procedure may be performed under ultrasound guidance - but this is not absolutely necessary.

1. Unless the patient is very sick, give IM pethidine half an hour before the procedure.
2. Prop the patient up in bed at 60 degrees.
3. An assistant should hold the child's arms and legs.
4. Scrub your hands, and put on sterile gloves.
5. Swab an area of 10 cm radius from the xiphisternum with iodine.
6. Inject 2 ml of 1% plain lignocaine into the skin and subcutaneous tissue at the left xiphicostal angle.
7. Aspiration is performed using the biggest Dwellcath available (14-19). The needle is inserted at the left xiphicostal angle and pushed slowly towards the left shoulder, keeping close to the chest wall.



8. Advance the needle slowly until fluid enters the syringe - this is usually at least 2 cm in. Slide the Dwellcath cannula gently further in at the same time as removing the needle and quickly connect a 20 ml syringe and 3-way tap.
9. SLOWLY aspirate the fluid into the syringe. If this is done rapidly, it may cause shock. When the syringe is full, turn the 3-way tap so that the side arm is open. Put 10 ml of fluid in a sterile bottle for bacterial culture, ZN stain for AFB, and protein. Discard the rest of the fluid into a container through a giving-set with the end cut off.
10. Turn the 3-way tap back again, and aspirate again until no more fluid is aspirated.
11. Remove the Dwellcath and clean off the iodine.
12. The dangers of pericardiocentesis include sudden death (from ventricular fibrillation), puncture of the heart, coronary artery or internal mammary artery, and pericardial shock if fluid is removed too rapidly.

Notes

1. If thick pus is obtained it will be necessary to drain the pericardium. This can be done surgically or through a lavage technique. Antibiotic therapy with cloxacillin should be continued for a minimum of 4 weeks.
2. Tuberculous effusions may be serous or fibrous and haemorrhagic. If the latter, surgical drainage through a large pericardial window is required to prevent the subsequent development of constrictive pericarditis. Tuberculosis treatment should be started immediately.
3. In a patient with a history and CXR suggestive of TB and with a small or moderate pericardial effusion which is completely black on ultrasound (serous effusion) and with no signs of tamponade, antituberculous therapy and steroids may result in rapid resolution of the effusion.

REFERENCES

- Hugo-Hamman CT et al. *Pediatr Infect Dis J* 13(1):13-18,1994. Tuberculous pericarditis in children: a review of 44 cases.
- Smith F. *Trop Doctor* 19(3):105-108,1989. Acute pericarditis with effusion and the general surgeon.

PERITONEAL DIALYSIS

This should only be used for acute, reversible renal failure.

1. Empty the bladder with a No.5 or No.8 feeding tube, unless it is certain that the bladder has just been emptied.
2. A sterile technique **MUST** be used.
3. Add 1000 u heparin to each litre of standard (1.6% dextrose) dialysis fluid.
4. Add potassium chloride 4 mEq (1.25 ml of 2 g in 8 ml solution) to each litre of dialysis fluid if the serum potassium is less than 4 mEq/l.
5. Swab the skin with iodine. Put on a cap, mask, sterile gown and gloves. Using a sterile technique, anaesthetise the skin and subcutaneous tissue with 2% plain xylocaine in the midline 2 cm below the umbilicus (ensure that the liver and spleen are not enlarged).
6. Before inserting the catheter, infuse 25 ml/kg of warmed dialysis solution into the peritoneal cavity over 15 minutes via a Dwellcath or Medicut.
7. Insert the peritoneal dialysis trocar and catheter through the anaesthetised area. After removing the trocar, direct the catheter downward into the pelvic cavity (for small children it may be necessary to cut off part of the catheter).
8. Establish that fluid flows freely from the catheter, then begin the dialysis. Use 50 ml/kg (maximum 500 ml) for each cycle. Run it in over 10 minutes, equilibrate for 30 minutes, and drain for 20 minutes. The peritoneal cavity must be drained completely with each cycle; this may require positioning the patient. The dialysis fluid must be warm.
9. An **ACCURATE** cumulative record of the volume of fluid infused and withdrawn **MUST** be kept. If a progressive positive balance develops, hyper tonic (7% dextrose) dialysis fluid can be used. If an excessive negative balance develops, the abdominal cavity should still be drained completely each cycle, but replacement can be made by intravenous fluids.
10. Record pulse and BP 4 hourly, and weigh daily.
11. If the blood glucose exceeds 10 mmol/l, give 0.1 to 0.2 u/kg of plain insulin IV.
12. Dialysis usually lasts about 48 hours. The main danger is from infection (causing peritonitis). In some patients the dialysis catheter blocks and has to be resited or replaced.

REFERENCE

Kempe CH, Silver HK, O'Brien D. Current pediatric diagnosis and treatment, Lange, 1978, p.508-9.

PERITONEAL TAP - DIAGNOSTIC

This may be used to detect the presence of bowel contents, pus or blood in the peritoneal cavity without resorting to formal laparotomy.

The usefulness of the procedure is limited by the fact that a negative tap does not absolutely exclude the presence of these fluids in the peritoneal cavity.

NEVER attempt diagnostic peritoneal tap in a patient who has had a laparotomy, because of the possibility of adhesions and penetration of the bowel with the needle.

1. The site of aspiration is usually to the left or right of the rectus muscle at the level of the umbilicus. Check that there is no hepatosplenomegaly or other abdominal mass in the area and that the bladder is empty.
2. Have the patient supine, but rolled slightly toward the side of the aspiration.
3. Clean the skin over a 10 cm radius with iodine.
4. Draw up 3 ml of sterile saline in a 10 ml syringe with a 21 gauge disposable needle on it.
5. Push the needle through the skin. Then start injecting the saline and push the needle into the peritoneal cavity. Then attempt to aspirate peritoneal fluid. Three attempts are usually made before the tap is said to be negative.
6. Alternatively, a nick is made in anaesthetised skin and a sterile trocar and cannula inserted into the peritoneal cavity. A small plastic catheter (or an infant feeding tube) can then be passed through the cannula into the upper and then the lower abdomen.
7. If any blood, bile, faeces or urine is aspirated, the result of the tap is positive and laparotomy will be required.
8. If the tap is negative, 10 ml/kg of normal saline can be instilled into the peritoneal cavity and aspiration attempted again.

REFERENCE

Radford A. PNG Med J 10:104,1967. Peritoneal tap.