CHAPTER 34

Acute Flaccid Paralysis

Acute flaccid paralysis is defined as weakness in one or more limbs, or the respiratory or bulbar muscles, resulting from damaged lower motor neurones. Poliomyelitis was the most important cause, but since it has declined other causes have become more important.

Classically, in acute flaccid paralysis there is weakness with reduced tone (flaccid weakness) and reduced or absent reflexes. Differentiating from upper motor neurone weakness is usually straightforward, but it should be remembered that acute spinal shock (e.g. caused by trauma) can initially cause flaccid paralysis before spasticity develops.

Pathophysiology and clinical presentations

Broadly speaking, there are two pathophysiological processes that cause acute flaccid paralysis (Fig. 34.1). These are direct viral damage of lower motor neurone cell bodies in the anterior horn of the spinal cord (e.g. polio, other enteroviruses, flaviviruses); and a para- or post-infectious immunologically mediated process damaging the motor nerves, and often sensory nerves (e.g. Guillain–Barré syndrome), sometimes caused by antibodies directed against the gangliosides (glycolipids in the nerve cell membranes). Recognizing the clinical features of these two patterns helps in determining the likely cause (Table 34.1).

Anterior horn cell damage causing acute flaccid paralysis

Polio

Infection with this enterovirus can be asymptomatic, can cause a mild non-specific febrile illness, viral meningitis or paralytic poliomyelitis, which can be spinal or bulbar. Paralytic poliomyelitis is biphasic, with a non-specific fever followed by a brief afebrile period before the central nervous system (CNS) is invaded. This is heralded by further fever and an acute-onset asymmetrical flaccid paralysis of one or more limbs, which may be painful. Since the World Health Organization campaign to eradicate polio using the oral polio vaccine, the number of cases has dropped from more than 350 000 cases in 1988 to approximately 1900 cases in 2002. Most of these came from South Asia (India, Pakistan and Afghanistan), West Africa (mainly Nigeria) and Central Africa (mainly Democratic Republic of Congo).

Enterovirus 71

Enterovirus 71 has caused epidemics of acute flaccid paralysis in recent years (especially in Asia), often in association with hand, foot and mouth disease. Many other enterovirus, Coxsackie virus and echovirus serotypes occasionally cause acute flaccid paralysis.

Japanese encephalitis virus

Japanese encephalitis virus, West Nile and other flaviviruses typically cause meningoencephalitis,

CAUSES OF ACUTE FLACCID PARALYSIS

	Direct viral damage to anterior horn cells (e.g. polio)	Immune-mediated damage to peripheral nerves (e.g. Guillain–Barré syndrome)
Paralysis onset	During (or straight after) febrile illness	Several weeks after febrile illness
Pattern of paralysis	Asymmetrical	Symmetrical
Time to reach maximum weakness	Short (e.g. 2–3 days)	Long (e.g. 7–14 days)
Sensory involvement	No	Often (depending on exact disease)
CSF	Increased lymphocytes (e.g. 100/mm^3)	Increased protein (e.g. 100 mg/dL, especially late in the disease)
Pain	Often limb muscle pain	Often back pain

Table 34.1 Clinical features to distinguish causes of acute flaccid paralysis.

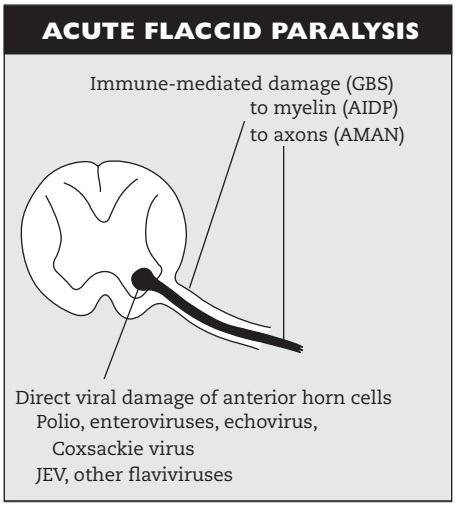

Fig. 34.1 Pathophysiology of acute flaccid paralysis. Immune-mediated Guillain–Barré syndrome (GBS) occurs in two forms: in acute inflammatory demyelinating polyneuropathy (AIDP) the myelin is damaged; in acute motor axonal neuropathy (AMAN) the motor axons are targeted. Viruses such as polio and Japanese encephalitis virus (JEV) cause paralysis by directly attacking the lower motor neurones (the anterior horn cells).

but flaviviruses can also present with a pure flaccid paralysis that can be clinically similar to polio (Chapter 33).

Immune-mediated causes of acute flaccid paralysis

Guillain–Barré syndrome is now recognized as a group of disorders classified according to the predominant type of nerve injury (axonal or demyelinating) and the main nerve fibres involved (motor, sensory, cranial). Different antiganglioside antibodies are associated with different diseases.

Acute inflammatory demyelinating polyneuropathy (AIDP, or 'classical' Guillain–Barré syndrome)

This typically presents several weeks after a febrile illness with back pain, then symmetrical ascending flaccid paralysis and sensory changes. Recovery is usual. Treat rapidly progressing symptoms with intravenous immunoglobulin if available.

Acute motor axonal neuropathy (AMAN, or Chinese paralytic syndrome)

This typically follows diarrhoea caused by *Campylobacter jejuni*. Symmetrical weakness is present with no sensory changes. Residual weakness is common. Occurs in summer epidemics in China.

Rare causes

Consider rare causes of paralysis if the diagnosis is not apparent.

- Any exposure to toxins?
- Any tick bites (tick paralysis is a slowly ascending paralysis that recovers when the tick is removed)?
- Exposure to rabid animal (paralytic rabies)?
- History of a severe sore throat with neck swelling (diphtheritic neuropathy)?
- Consumption of poorly preserved food (botulinum toxin)?

Nerve conduction studies

Where available, nerve conduction studies may help distinguish further:

- *Anterior horn cell damage* — motor amplitude is reduced because motor cell bodies have been damaged.
- *Classical Guillain–Barré syndrome* (autoimmune demyelinating polyneuropathy) — motor and sensory nerves have reduced conduction velocities and delayed distal latencies because demyelinated nerves conduct more slowly.
- *AMAN* (Chinese paralytic syndrome) — motor amplitudes are reduced because motor axons have been damaged.

Further reading

Anonymous. Progress toward global poliomyelitis eradication, 1999. *Morbid Mortal Week Rep* 2000; **49**: 349–54.

Bolton CF. The changing concepts of Guillain–Barré syndrome. *New Engl J Med* 1995; **333**: 1415–16. [Good explanation of acute motor axonal neuropathy as a form of Guillain–Barré syndrome.]

Solomon T, Willison H. Infectious causes of acute flaccid paralysis. *Curr Opin Infect Dis* 2003; **16**: 375–9. [Annotated review of main infectious causes.]