Differential diagnosis of acute flaccid paralysis: Poliomyelitis in developing countries

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The WHO and Indian guidelines for the diagnosis of paralytic poliomyelitis emphasize the asymmetry observed in paralysis as the main diagnostic feature, without making clear whether the observed asymmetry refers to limb or muscle. However, in developing countries most children with fever are given one or more unnecessary intramuscular injections. These injections subsequently determine which limbs and muscles are paralysed. Indian children with acute poliomyelitis have been analysed by site of injections received: more than 50% children showed symmetry of paralysis by limb, i.e. both the limbs were paralysed, rising to more than 70% among those children who had received injections in both legs. One quarter of the children showed symmetrical paralysis of muscles, rising to one half among children who had received injections in both the legs. As there have been no follow-up studies at 60 days after paralysis. I have used records from rehabilitation clinics in Pakistan, where the affected children showed severe paralysis of the distal muscles. The WHO and Indian Government guidelines suggest that symmetrical paralysis is caused by Guillain-Barré syndrome (GBS) or transverse myelitis (TM), and rule out polio. However, the most important determinant of symmetry, severity, and location of paralysis is the number and site of injections received. The official guidelines are incomplete, unclear and misleading for use with affected children, most of whom will have received injections prior to paralysis. The official forms should be amended to include details of injections and muscle paralysis.

NATIONAL IMMUNIZATION DAYS (NIDs) may reduce the incidence of poliomyelitis, but not eradicate it. To ensure eradication there must be surveillance, identification of cases, the sending of faecal samples for laboratory investigations, and examination for residual paralysis after 60 days. To prepare for eradication, WHO produced guidelines[1] to distinguish polio from other causes of acute flaccid paralysis (AFP). If faecal samples are not collected or yield no virus, diagnosis will be based on these guidelines. However, initial diagnosis of Guillain-Barré syndrome (GBS), transverse myelitis (TM), etc might preclude collection of faecal samples. Even in European countries, it is difficult to get doctors to collect faecal samples from cases of AFP.

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India, with the most cases of polio, has its own guidelines[2]. Both guides emphasize that polio paralysis in developing countries affects the very young, and is asymmetric. It is therefore important that this criteria hold true for cases from the area.

What the guidelines say:

The asymmetry of paralytic polio is stressed in the WHO manual[1]: 'anatomic distribution of weakness is usually asymmetric, beginning in lower or, less often, upper extremities (p. 6)', 'Symmetry uncommon' (Table 1), and 'characteristically asymmetric and may be localized, such as to a single limb' (Appendix III, detailed clinical criteria).

The Indian Field Guide[2] also stresses the asymmetry of poliomyelitis: 'presents as asymmetrical lower motor neurone flaccid paralysis', 'Main signs: asymmetrical paralysis'. 'As it is asymmetrical patchy paralysis, muscle strength varies in different groups of different limbs. However, proximal muscle groups are more involved as compared with distal ones' (p. 19).

'Flaccidity: acute, asymetrical, proximal (polio); acute, symmetrical, distal (GBS); acute, lower limbs, symmetrical (TM)' (Table p. 22).

Both manuals emphasize that the important feature of polio is the asymmetry of paralysis. Muscles are mentioned, but on the forms there is no space for recording paralysis of individual muscles or of asymmetry. In the guidelines, symmetry of paralysis, however, defines both GBS and TM. It is not clear whether symmetry is based on limb symmetry, i.e. both legs or both arms paralysed, or whether the symmetry is based on the severity of paralysis of particular muscles in each limb, e.g. muscle strengths of 2 (MRC scale, see below) in the gluteus, quadriceps and ankles in both legs. Nor is it clear whether severe paralysis in both legs would be judged as symmetrical even if different muscles were affected.

I have re-examined the records of muscle paralysis of 262 children at JIPMER in South India 1987–1990 with diagnosis of acute poliomyelitis following fever[3]: five case histories were unsuitable for this analysis. Paralyses of children attending two rehabilitation clinics and a polio camp in Pakistan have also been re-analysed[4] (Table 2). The children have been grouped according to their injection history, with symmetry of paralysis noted for muscles and limbs. Measuring severity of paralysis is difficult, particularly in very young children: the qualitative MRC scale uses 5 for normal and 0 for complete paralysis. I have ignored differences of a single grade where only one muscle is involved e.g. 1 in the right gluteus and 2 in the left.

The Indian manual suggests that 'A history of intramuscular infection (*sic*) in the gluteal region will favour the diagnosis of traumatic neuritis (TN)' (ref. 2). There is no evidence that intramuscular injections cause TN except in very exceptional cases: pediatricians, dentists

and anaesthetists all give injections in or near nerves without causing paralysis. Injections which damage the sciatic nerve would be expected to result in sensory as well as motor damage. If the sciatic nerve was damaged, one would expect a violent reflex response and a howl of pain. The nerves which supply the gluteus and quadriceps muscles leave the sciatic nerve before the site of injections. Therefore damage to the sciatic nerve should spare damage to those muscles. However, as the nerves diverge to supply either the gluteus or the quadricep, only one of these muscles should be paralysed. However, in India about 70% or more of the hospital cases of acute polio have received one or more injections in one or more limbs[5], and paralysis is related to the number and location of the injections, both in severity of muscle weakness and in the pattern of paralysis[3]. Almost all the injections are given for fever within 48 h of onset of paralysis.

If symmetry is defined by limb, 52% of the children (Table 1) would have been considered by the WHO and Indian manuals as likely to be cases of GBS or TM rather than polio. However, since 13 of these children died, they would therefore be considered as polio cases (see below), reducing the remainder to 47%. The proportion with symmetry differed from 40% to 72% according to the number and site of injections received. Nearly three quarters of the children who received injections in both legs were paralysed in both legs, and would have therefore been presumed to have GBS or TM rather than polio.

If symmetry is defined by muscle strength, then 25% of the children showed symmetrical paralysis (Table 1). In this group nine children died, reducing to 21% those with symmetry and unlikely to be considered polio cases. Again, the proportion of children presumed not to be polio cases ranged from 17% to 50% according to the number of injections. A total of 390 paralysed legs were analysed further, in which 23% were cases with both legs totally paralysed and another 21% were single legs with total paralysis. In 23% of the legs, paralysis of hip, knee and ankle was equal. In 28% of the legs, the hip and knee were more paralysed than the ankle and in only 6% was the ankle more paralysed. In general, toes were spared, although paralysis was difficult to determine. Thus, involvement of the gluteus and quadriceps was maximal. In cases of aggravation (muscle injected < 48 h before paralysis), the injected limb may be spared, although paralysis may occur in other limbs. For paralysis to occur, there must be enough poliovirus already in the spinal cord.

Death after fever and paralysis brings a retrospective WHO diagnosis of polio. In my sample of 257, fourteen children were diagnosed on death as polio cases, regardless of the symmetry of their paralysis (see above). However, if because of their symmetrical paralysis, they were initially not considered as polio cases, stool specimens might not have been taken and sent for analysis, thus missing the opportunity to isolate poliovirus. Fatal cases were over-represented in our sample, because their records were more complete. The Indian Manual says

Table 1. Symmetry of paralysis of acute poliomyelitis in children at JIPMER, Pondicherry, India (reworking of original data used for ref. 3)

	Injections					
	None	1 leg	2 legs	Site and no. not known	Provocation	All
N	71	65	40	69	12	257
% Symmetric by limb	46	40	72	58	50	52
% Symmetric by muscles	21	17	50	22	25	25

Injections were given less than 48 h before onset of paralysis (aggravation) except in the case of provocation when injections had been given 5 to 30 days before paralysis.
For 69 children, the number and location of injections were not known.

Table 2. Symmetry of residual paralysis of poliomyelitis in children attending clinics at Peshawar and a polio camp at Kohat, Pakistan

	Injections					
	None	1 leg	2 legs	Other	Forms incomplete or cannot remember	All
N	29	102	67	24	31	253
% Symmetric by limb	34	10	84	36	32	37
% Symmetric by muscles	17	4	51	33	16	22

There were 40 children attending a polio camp at Kohat or a rehabilitation clinic in Peshawar and 213 attending another clinic in Peshawar[4]. Although all had suffered paralysis as children, the ages when seen, ranged from 1 to 20 years.

that 'In 2 to 20% of the cases the outcome may be fatal'[2]. However, the case-fatality rate was 1.85% for 175,081 children with acute polio seen in sentinel hospitals 1976 to 1984 (ref. 6): higher rates were based on severe cases admitted to specialist hospitals[5].

At least 76% of the children in Pakistan had received an injection prior to paralysis (Table 2) and was 87% for those with completed forms. The patterns of symmetrical paralysis were similar to those from India (Table 1). However, the biggest difference was the absence of paralysis in the uninjected leg: paralysis in legs without injections is less severe and muscles may recover[3,5]. Unlike the Indian children with acute paralysis, the Pakistani children had more severe paralysis in the quadriceps than the gluteus and even more severe paralysis in the ankle (average strengths 1.9 gluteus, 0.7 quadriceps and 0.3 ankle in injected legs, MRC qualitative scale): paralysis was more severe in distal muscles.

In conclusion, the major factor determining the location and severity of paralysis in polio is the number, timing and location of injections[3]. The EPI Poliomyelitis Case Investigation Form (Appendix I of ref. 1) has no mention of injection. The Indian Case Investigation Form[2] asks 'Any injections during 30 days before paralysis onset: Yes/No/Unknown'. This is not good enough: the effects of provocation (injections 6–21 days before paralysis) and aggravation (injections < 48 h before paralysis) are different. It is important to know when, how many and where and what has been injected. Unfortunately, few parents know what was injected[5].

Surveillance for residual paralysis at 60 days although crucial for eradication, is not easy in India and I know of no follow-up studies of muscle paralysis after 60 days (see bibliography S71–S98, ref. 5). In 1995, there were 171 cases of polio among residents of Mumbai (Bombay), another 92 among non-residents, of whom 26 were out of state[7]. Some of the 66 cases from Maharashtra might be traceable, but the 22 from Uttar Pradesh would be from poor and illiterate families who would return to their villages more than 1500 km away. Clearly, there was a very high rate of polio among these migrants who had missed immunization in their home states and also in Mumbai. As follow-up studies are difficult, it is even more important that the initial diagnosis be accurate.

Polio paralysis in the Indian sub-continent and probably Africa, differs from that of the mainly much older children in temperate countries. Major differences result from unnecessary injections given for fever caused by poliovirus already in the spinal cord (aggravation). The different diagnoses of the paralysis included in AFP should be changed to reflect reality. As Lambert *et al.*[8] reported, the '3 children (with paralytic poliomyelitis, Afghanistan) had received an injection before paralysis and would not be included if WHO definition was strictly applied'. The investigations and forms must report clearly the time, location, etc of injections and must include more details of muscle paralysis.

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