The Prevalence and Severity of Joint Problems and Disability in Patients with Poliomyelitis in Urban India

S. Baliga[*,1], T. Mcmillan[1], A. Sutherland[1] and D. Sharan[2]

[1]*University of Aberdeen, Aberdeen, Scotland*

[2]*Mobility India, Bangalore, India*

Abstract: Poliomyelitis is caused by an enterovirus infection of the anterior horn cells in the spinal cord. Up to 40% of survivors recover full muscle strength, however 60–90% are left with varying degrees of residual paralysis, where the patient suffers from cramping myalgia and lower motor neuron pattern weakness. This study aimed to identify and quantify, in terms of prevalence and severity of the types of joint deformities encountered in polio sufferers. It also aimed to assess the disability caused by such problems. Finally we documented the provision and use of mobility aids, orthotics and surgery in the patient group.

Impairment was confined to one lower limb, and this is consistent, as the majority of patients were infected in infancy. The study found that pes cavus, scoliosis, flexion deformity of the knee and true lower-limb shortening accounted for over half of the deformities found. The mean Barthel Disability score was 19 and over 80% of patients used at least one aid, usually in the form of a Knee-Ankle-Foot Orthosis (KAFO). Surgery also plays a large in role in the management of polio patients, however necessity needs to be assessed on an individual basis taking into account many aspects of the patient's life.

Keywords: Disability, joint deformity, polio rehabilitation, poliomyelitis.

BACKGROUND

Poliomyelitis is caused by an enterovirus infection of the anterior horn cells in the spinal cord [1]. Transmission occurs mainly through faeco-oral and after a seven-day incubation period, symptoms include fever, fatigue, headache, and pain in the limbs, the so-called 'pre-paralytic' stage. Children under five years are at the highest risk of contracting the illness, however the consequences of adult disease are generally worse.

Up to 40% of survivors recover full muscle strength; however 60–90% are left with varying degrees of residual paralysis [2], where the patient suffers from cramping myalgia and lower motor neuron pattern weakness.

Treatment is supportive, about 2-5% of children and 15-30% of adults with paralytic disease die, usually due to respiratory failure. Complete paralysis of the muscles around a joint may permit subluxation to occur. When opposing muscle groups are unequally affected, contractures are apt to occur in the stronger muscles, restricting movement of the joint.

The residual effects can lead to severe physical impairment, which in turn leads to disability, which may render the patient incapable of carrying out social functions.

At its annual meeting in Geneva in May 1988, the World Health Assembly (the governing body of the World Health Organisation), resolved to eradicate polio from the world by the year 2000. Since the Global Polio Eradication Initiative (GPEI) was launched, the number of cases have decreased by over 99%, from an estimated 350,000 to 1919 in 2002. In the same time period, the number of polio-infected countries was reduced from 125 to 7 [3].

Despite these achievements, the Polio Eradication Initiative faced an increase in global cases over 2001, 1919 cases were reported (as of 16 April 2003), compared to 483 in 2001. This increase can be attributed to epidemics in India and Nigeria. WHO had described India as the 'highest priority country', due to having the majority (83%) of the world's cases in 2002. In 2014, India celebrated three years without reporting any case of Polio. Yet, an estimated 20 million people continue to live with post polio sequelae worldwide [4]. Only Pakistan, Nigeria and Afghanistan have never interrupted wild poliovirus transmissions, remaining in endemic areas.

AIMS AND METHODS

This study aims to identify and quantify, in terms of the prevalence and severity of the types of joint deformities encountered in polio sufferers. It also aimed to assess the disability caused by such problems. Finally we aimed to document the provision and use of mobility aids, orthotics and surgery in the patient group.

Data collection was performed in 2005. All the participants in the study were recruited through Mobility India and lived in the city of Bangalore. Mobility India is an organization that works in the field of rehabilitation,

*Address correspondence to this author at the Department of Trauma and Orthopeadics Aberdeen Royal Infirmary, Foresterhill Road, Aberdeen. AB25 2ZN, Scotland; Tel: +447815676059;
E-mail: santoshbaliga@doctors.org.uk

1874-3250/15

primarily for people with locomotor disabilities. Thirty patients were chosen at random by community project workers, and were visited in the community.

Information collected from every patient included name, date of birth, sex, onset of polio paralysis, braces, aids or prostheses used, past surgical procedures, co-morbid conditions (especially pertaining to musculoskeletal problems) and the place of visit.

Examination of all limbs included: briefly looking for wasting, loss of tone and diminished reflexes to sustain the diagnosis of poliomyelitis. A more detailed assessment involved measurement of power using the Medical Research Council (MRC) grading of power, and range of movement in all major joints, using a goniometer. As well as this, true limb-length and Thomas's test for fixed flexion deformities of the hip were performed. The back was then examined to look for spinal deformities.

An assessment of disability was made using a Barthel's Index of Activities of Daily Living [5]. Notes held on every patient by Mobility India were then consulted to clarify details of past surgical procedures.

RESULTS

Population

Data from 26 patients was collected. One other patient's data, which was thought to have polio, was discounted, as the pattern of impairment did not fit that of poliomyelitis.

The group consisted of 13 males and 13 females, ages ranged from 7 to 39, (mean 18.9).

Age of onset ranged from 3 months to 8 years, the mean age of onset was 1.3 years in males, 2.2years in females and 1.7 years overall.

Joint Deformity and Impairment

Fourteen of the patients were impaired in only one lower limb; 10 patients had significant bilateral lower-limb impairment. One patient had weakness of one lower and one ipsilateral upper limb and one patient had severe paralysis of all four limbs.

The most common deformity was pes-cavus, found in 20 patients followed by scoliosis, flexion deformity of the knee (Fig. **1**) and true lower limb shortening (Fig. **2**). Table **1** summaries all skeletal deformities found.

Scoliotic deformities were all confined to the thoracolumbar region; 14 out of 16 of the patients had a 'C' shaped scoliosis. One patient suffered from the double curve deformity.

The lower-limb was the site for the vast majority of problems; only one patient had deformities of the upper limb, however four other patients had some upper limb weakness.

The severities of Hip and Knee deformities are outlined in the Table **2**. True lower-limb shortening was on average around 2.5cm. Although shortening was mostly confined to the paralysed limbs, one patient had a shortening of a clinically 'normal' left lower limb.

Table 1. Deformities.

Deformity	Number of Patients Affected	Number of Patients Affected Bilaterally
Pes-Cavus	20	2
Scoliosis	16	-
Flexion of Knee	14	1
True lower limb shortening	11	0
Flexion of Hip	7	1
Genu-Valgum	5	0
Flexion of Toes	5	0
Genu-Recurvatum	4	2
Varus of Ankle	4	2
Valgus of ankle	4	1
Pes-Planus	2	0
Eqinus Ankle	1	0
Lordosis	1	-
Flexion of Elbow	1	0
Claw Hand	1	0

Fig. (1). Flexion deformity of knee.

Fig. (2). True limb shortening.

Table 2. Severity of hip and knee deformities.

Deformity	Average Severity (°)	Ratio of Male: Female Suffers
Hip Fixed Flexion	15	6:1
Fixed Knee Flexion	27	6:7

Orthotics and Mobility Aids

Over 80% of patients used at least one aid, usually in the form of an orthotic but other devises included, elbow crutches, armpit crutches, raised shoes and wheelchairs. The use of mobility aids and orthotics is summarized in Table **3**. The most commonly used orthotic was the Knee-Ankle-Foot Orthosis (KAFO); this was followed by the Ankle-Foot Orthosis (AFO). A KAFO and AFO are shown in Fig. (**3**). None of the patients used a Hip-Knee-Foot Orthosis or a spinal brace.

Table 3. Use of orthotics and mobility aids.

Aid	Left	Right	Bilateral	Total
KAFO	8	7	2	17
AFO	1	3	0	4
Elbow Crutches	1	0	2	3
Raised Shoe	1	1	0	0
Wheelchair	-	-	-	1
Armpit Crutches	-	-	-	1

Fig. (3). Right sided ankle foot orthosis (AFO) and left sided knee-ankle foot orthosis (KAFO).

Surgery

Ten patients had undergone surgery, all for lower-limb contractures, three male and seven female. In addition to this, one male patient had refused surgical correction for knee flexion deformity and one patient had had surgery of an affected limb after trauma. No patients had had any upper-limb or spinal surgery.

The average age of surgery was 12.9 years, females tended to have surgical intervention at an earlier age, their mean age being 8.8 years compared to males with a mean age of 17years.

In all but one case there was little record of the severity of contractures before surgery. Out of the five patients that had undergone hip contracture release surgery, only one had a residual 10° flexion deformity. Out of the eight that had undergone knee contracture release, six had a recurrence or residual degree of contracture, averaging around 12°. Out of the six patients that had undergone ankle surgery (all for equinus deformity) none had recurrence of the deformity.

Disability

The mean, mode and median Barthel score was 19. The females scored marginally higher with 19.1 compared to the male's average score of 18.9. Scores ranged from 14 to 20. A breakdown of these scores is shown in Table **4**. The most commonly encountered difficulty for patients seemed to be stairs. Other common problems included mobility and transfer.

Table 4. Patient barthel scores.

Barthel ADL Score (/20)	14	15	16	17	18	19	20
Number of Patients	1	0	0	1	3	11	10

DISCUSSION

Population

All patients included in the study reported a history of a few days of a febrile illness in childhood, followed by paralysis. In most there was some resolution, however all had residual weakness. All patients exhibited signs of atrophic flaccid paralysis with diminished reflexes but integral sensation. These patterns are consistent with that seen of poliomyelitis.

No patients in the study group were younger than seven years old; this perhaps outlines the success of the polio eradication at least in the State of Karnataka. As Table **5** shows, there have been 36 cases in the five years prior to data collection, all of which have been confined to the northern part of the state. In fact the city of Bangalore itself, had last seen a new case of polio in 1999. It was thus unlikely to find patients younger than this.

Deformity

Impairment was confined to one lower limb, and this is consistent, as the majority of patients were infected in infancy. Had the average age of onset been over 5 or 15

years of age, there would have been a greater proportion of patients with paraplegia and quadriplegia.

Table 5. Data compiled from WHO SEAR polio bulletin [6] and a blot on polio eradication drive, DH news service [7].

Year	Incidence in India	Incidence in Karnataka State
2000	265	1
2001	226	0
2002	1600	0
2003	225	34
2004	46	1

The most common deformity was pes cavus and not flexion deformity of the knee as reported in other literature, this is partly because of surgical contracture release and partly due to the high uptake of calipers, in particular the KAFO, which has allowed many patients to walk and hence allowed them to use muscle groups effectively. This has prevented muscle shortening and thus reduced the incidence of contractures.

Out of the 22 pes cavus deformities seen, 20 had grade III weakness or below of plantar flexion, 17 had weakness of hallucis extension. This pattern is indicative of L5 and S1 radicular involvement. In 14 out of the 22 cases, big toe flexion was clinically stronger than toe extension.

These results highlight the pathogenesis of pes cavus deformity, where the anterior horn cells at S2-4 supplying the intrinsic muscles of the foot are less damaged than cells at L5 and S1. The mean length of disease in the pes cavus group was 17.5 years, in comparison to the unaffected group, which was 7.5years. This suggests that this pes cavus is more is more likely to arise as time goes by.

Scoliosis was the second commonest deformity, being present in over 60% of patients to some degree. Curvatures secondary to poliomyelitis take one of two forms: a curve resulting from asymmetric paralysis or a collapsing spine, involving almost the whole trunk, due to symmetric paralysis [8]. Leong *et al*. claimed that because many patients have limb involvement as well spinal deformity, bracing tends to be poorly tolerated. This perhaps explains why no patients were fitted with spinal orthosis.

Limb shortening is common in poliomyelitis. Occasionally limb lengthening can initially occur after paralysis, but after a few months gradually increasing shortening is the rule in the growing child. Shortening of the lower limb may cause the patient to walk with a more severe limp than would otherwise be the case [3]. Although 11 patients had a true lower limb shortening only two used a raised shoe. This disparity could be that the majority of patients had shortenings of less than 2.5cm, which are generally well tolerated. Raised shoes were used in patients who's apparent shortening was over 4cm.

Aids

It is not surprising perhaps that the most common aid used by far was the KAFO. This is directly linked to the high incidence of lower limb impairment. All patients in this group had grade III or less weakness of knee movement. The knee joint, relying heavily on its muscles for stability, becomes unstable and unable to support weight during walking. A KAFO maintains the knee in the extended position allowing the affected lower limb to be one solid column, capable of supporting body weight. It is worth noting that all patients using KAFOs rated their mobility as 3/3 in comparison with the rest of the patients whom averaged a mobility score of only 2.4. The main cause of disability in this group of patients arose in climbing stairs, 11 of the 17 patients needed some amount of help.

The second commonest aid was the AFO; all the patients in this group had weakness of muscles supplying the ankle, and hence needed support to stability for walking. Apart from muscular weakness ankle instability can also arise from bony deformity.

Bilateral elbow and armpit crutches in combination with KAFOs were principally utilized by patients with substantial weakness in both lower limbs. Crutches provided a point of stability from where they could effectively use their existing muscle power to transfer and move. One patient having paralysis affecting all four limbs was dependent on the use of a wheelchair for mobility around the local area.

Surgery

Surgery in poliomyelitis aims to correct limb deformity, improve function and stabilize the limb or spine. Previously it was to eliminate braces, but now the goal is to improve bracing options. All patients in the study underwent tendon release surgery. Ilio-tibial, hamstring and Achilles release were performed for hip, knee and equinus contractures respectively.

Table 6. Comparing rates of operative procedures.

Operation	% of Patients Recommended for Surgery by Perry *et al.*	% of Patients in Study
Ilio-tibial lengthening	4.5	19
Hamstring release	<4.5	30
Achilles tendon release	12.5	23

The most common type as reported by Perry *et al.* [9] in their group of patients was Achilles' tendon release, recommended in 12.5% of their patients, however in this study the most common operation was hamstring release for flexion deformity of the knee, performed on 30% of patients. In fact there was a higher percentage operated on for all types of surgeries. These figures are summarized in Table **6**.

Surgery in general being reserved for hip and knee contractures of over 30°, perhaps suggests that the patients in

this study were more severely affected than those treated by Perry *et al*. This could be due to infection with different poliovirus types, poorer access to health care in particular physiotherapy or due to a bias in the population.

Although not well documented in case notes, patients themselves reported vast reductions in the severity of contracture after surgical procedures. However one patient needed further contracture release operations after recurrence of deformity ten years after. The patient had failed to carry out her daily stretches as recommended by physiotherapists.

There were no documented tendon transfers, arthroplastys or arthrodesis; this is probably because of the relatively young age group of patients, who had little joint pain or joint degeneration.

Disability

The mean age of patients was 18.9 years: It may be that sufferers from polio have a shorter life expectancy and that many of the older patients have passed away. It may also be that the older patients have moved away from the city, to rural areas where the cost of living is much less. An alternative explanation perhaps is that the organisation attracts younger patients. Whereas older patients whom have lived for over forty years and have managed to find a way of living and working with their impairment, younger patients feel the need to completely overcome their handicap and hence have equal opportunities in education and work.

The extent of their success can perhaps be partially estimated by Barthel Disability Scores. A mean of 19 through the study group, would suggest that these patients are coping surprisingly well, the majority being independent in all aspects of personal care. This could be down to the good utilisation of aids, which have allowed most patients to live normal lives despite the severity of their impairment.

Limitations

It is unfortunately common, that within the poor and uneducated population many problems are labeled as 'polio'. Thus it was felt import to exclude differential diagnoses. These include peripheral neuropathies such as lead poisoning, Charcot-Marie-Tooth syndrome and Guillain-Barre syndrome as well as Motor Neurone Disease (in particular Progressive Muscular Atrophy type) and Cerebral Palsy. A thorough history and examination was the tool used to exclude any dubious diagnoses.

One of the patients seen was a 23 year old female, who had been diagnosed with 'polio' at the age of four months by an unknown health professional. She herself was unable to give a first hand account and reported what she had been told by her mother. Examination revealed a spastic marked weak right lower limb, with increased reflexes and loss in sensation. She claimed ever since she could remember she never had a loss of sensation in the affected limb. Data collected from this patient were discounted as the diagnosis of polio was not clear-cut.

Full muscle strength varying with sex and age made the job of objectively MRC scoring particularly tricky. Where possible, 'full power' was judged to be the unaffected opposite limb. However studies by Beasley [10] over 30 years ago showed that strength rated grade five on the basis of manual testing in patients who had poliomyelitis was equivalent to 53% of the strength in normal controls. This backed by Bodain [11] who, in a study of acute poliomyelitis, found that the virus had invaded 97% of anterior horn cells, on average 47% of them recovered.

Visits in the community were particularly challenging, crowded and cramped conditions made some aspects of examination very difficult. Due to inadequate facilities such as a clean flat bed, Thomas's test was not performed on two patients.

The Barthel ADL scores may underestimate the true problems as many of the patients may have been slow to admit to problems of incontinence, especially in circumstances where there was little privacy. Another problem with the Barthel scale to be noted is that scores are sometimes difficult to assess consistently. Because environment plays such a large role in disability, patients who score poorly in one particular environment may well score better in another. For example many patients claimed that toileting was independent so long as there was a seated toilet. However some of the patients had only a squatting toilet at home and thus sometimes needed assistance.

CONCLUSION

After the launch of the Global Polio Eradication Initiative by World Health Organisation in May 1988 the number of new cases per year have decreased by over 99%, from an estimated 350,000 to 416 in 2013 [12]. The estimate incremental net benefits of the GPEI between 1988 and 2035 are 40-50 billion dollars, and low-income countries account for approximately 85% of the total net benefits generated by the GPEI [13]. Furthermore, Khan *et al*. predicted the program to prevent 855,000 deaths, 4 million paralysis cases and 40 million disability adjusted life years (DALYs) over the years 1970-2050 [14]. Although new cases of polio are thankfully rare, India still has almost 4 million victims with various types of musculoskeletal problems.

Impairment was confined to one lower limb, and this is consistent, as the majority of patients were infected in infancy. The study found that pes cavus, scoliosis, flexion deformity of the knee and true lower-limb shortening accounted for over half of the deformities found.

Over 80% of patients used at least one aid, usually in the form of a Knee-Ankle-Foot Orthosis (KAFO). This directly linked to the high incidence of lower limb impairment. All patients in this group had grade III or less weakness of knee movement. The joint relying heavily on its muscles for stability becomes unstable and unable to support weight during walking. A KAFO maintains the knee in the extended position allowing the affected lower limb to be one solid column, capable of supporting body weight. It is worth noting that all patients using KAFOs rated their mobility as 3/3 in comparison with the rest of the patients whom averaged a mobility score of only 2.4.

Surgery plays a large in role in the management of polio patients, it aims to correct limb deformity, improve function and stabilize the limb or spine. When successful it can subs-

tantially reduce impairment and hence improve disability, however, necessity needs to be assessed on an individual basis taking into account many aspects of the patient's life.

The mean Barthel Disability score was 19. The most commonly encountered difficulty for patients seemed to be stairs, many needing help in getting up and down. This would suggest that these patients are coping surprisingly well, the majority being independent in all aspects of personal care. This could be down to the good utilisation of aids and surgical procedures, which have allowed most patients to live normal lives despite the severity of their impairment.

Whilst GPEI aims to eradicate Polio it has also targeted residual paralysis and its surveillance, with the aim to have at least 80% of documented cases of acute flaccid paralysis having a follow-up examination 60 days after the onset of symptoms looking specifically looking for evidence of residual paralysis. This will in turn allow earlier patient identification, timely provision of treatment and support to maximize quality of life.

CONFLICT OF INTEREST

The authors confirm that this article content has no conflict of interest.

ACKNOWLEDGEMENTS

Declared none.

REFERENCES

[1] Corey L. Harrison's Principles of internal Medicine. In: Isselbacher K, Adams R, Braunnwald E, Eds. 14th ed. McGraw-Hill: USA 1999; pp. 812-3.

[2] Neumann DA. Polio: its impact on people of the United States and the emerging profession of physical therapy. J Orthop Sports Phys Ther 2004; 38(8): 479-92.

[3] Huckstep R. Poliomyelitis, A guide for developing countries-including appliances and rehabilitation. WorldOrtho Inc. 1997. Available on: www.worldortho.com

[4] Gonzalez H, Olsson T, Borg K. Management of post polio syndrome. Lancet Neurol 2010; 9(6): 634-42.

[5] Mahoney RI, Barthel DW. Functional evaluation: the Barthel index. Md State Med J 1965; 14: 61-5.

[6] SEAR Polio Bulletin. World Health Organization Regional Office for South-East Asia Poliomyelitis Surveillance: Weekly report for week 1, 2002. Vaccines and Other Biologicals 2002; 6(1).

[7] A Blot on Polio Eradication Drive. Decan Herald, DH News Service Bangalore, January 01, 2004.

[8] Leong JC, Wilding K, Mok K, Chow SP, Yau AC. Surgical treatment of scoliosis following Poliomyelitis: A review of one hundred and ten cases. J Bone Joint Surg Am 1981; 63(5): 726-40.

[9] Perry J, Keenan MA. Post polio corrective surgery: then and now. Post-polio health international. Available from: www.post-polio.org/educa.html

[10] Beasley WC. Quantative muscle testing: principles and applications to research and clinical services. Arch Phys Med Rehabil 1961; 42: 398-425.

[11] Bodain D. Motorneuron disease and recovery in experimental poliomyelitis. In: Halstead LS, Wiechers DO, Eds. The Late effects of Poliomyelitis. Miami: Symposia Foundation 1985; pp. 45-55.

[12] WHOPoliomyelitisFact Sheet No. 114 WHO, Geneva (2013). Avaiable from: http://www.who.int/mediacentre/factsheets/fs114/en/index.html

[13] Deintjer TR, Pallansch M, Cochi S, *et al*. Economic analysis of the global polio eradication initiative. Vaccine 2010; 29(2): 334-43.

[14] Khan M, Ehreth J. Costs and benefits of polio eradication: a long-run global perspective. Vaccine 2003; 21: 702-5.

Received: January 4, 2015 Revised: April 18, 2015 Accepted: April 26, 2015