Congenital Flaccid Bulbar Palsy

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Weakness of the muscles supplied by the bulbar nuclei may be congenital, producing feeding difficulties immediately after birth and dysarthria at a later date.

Most commonly the bulbar palsy is of supranuclear type, and forms part of the clinical picture of severe generalized cerebral palsy, usually of the athetoid variety. The defect may, however, be confined to the bulbar muscles. In these cases the infant is found to suck poorly in the neonatal period and to have difficulty in chewing and swallowing when weaned. Speech may be delayed and a characteristic dysarthria noted, so that the child may be incorrectly regarded as of subnormal intelligence. Drooling may be excessive, while chewing continues to be laboured. On examination the jaw-jerk is found to be exaggerated and the tongue to move stiffly.

Less often the congenital bulbar palsy is of lower-motor-neurone type. Here there is likely to be nasal regurgitation of milk in infancy and early childhood. Again sucking is often poor. Later these children find they cannot easily blow their nose or blow a musical instrument without the nostrils being pinched shut. Delay in speech is associated with a nasal dysarthria. Such patients evidently form a small proportion of those seen at speech-therapy clinics, in one of which Ingram (1959) found no lower-motor-neurone disorders in a series of 189 patients.

If the lower-motor-neurone paralysis is progressive the probability that the child is suffering from Werdnig-Hoffmann disease must be considered. By contrast, the following 10 cases form a moderately homogeneous group in which a *non-progressive* flaccid bulbar palsy was directly responsible for the presenting complaint. It will be seen that, as in those children with supranuclear lesions, the bulbar palsy may be only one feature of severe generalized but, in this case, flaccid paralysis. Similar cases have been described previously—for example, Benson (1962)—but only as sporadic single case reports. Congenital flaccid bulbar palsy is well recognized as a feature of Möbius's syndrome or facial diplegia (Möbius, 1892), but in published descriptions of this malady (Henderson, 1939 ; Evans, 1955) bulbar symptomatology is never prominent.

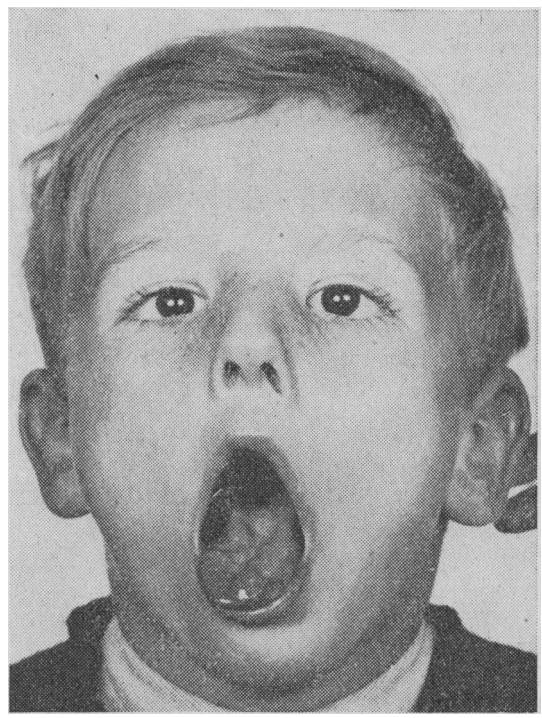
All the cases described below were under the care of Dr. P. H. Sandifer unless otherwise stated.

Case Summaries

Case 1.—Girl. Pregnancy complicated by marked hydramnios. Normal delivery. Birth weight 8 lb. 4 oz. (3,740 g.). Right talipes equinovarus. Cyanosed at birth for three minutes. Did not suck well. Slow feeder, milk regurgitated through nose; failed to swallow saliva. Required tube feeding. Admitted Hospital for Sick Children (Sir Wilfrid Sheldon) at 7 months. On examination: pale, floppy baby, dribbling from mouth ; bilateral facial paresis ; immobile palate ; generalized hypotonia, but no other neurological signs ; right talipes. Laryngoscopy (Mr. J. Crooks): " mouth, tongue, and pharynx formation normal, but movements of muscles poor. Larynx normal. Entrance to oesophagus normal, no stricture present. Oesophagus wide and lax." Propyliodone (" dionsil ") swallow: medium passed freely into the trachea via the glottis. Very little passed into the oesophagus, and no definite swallowing seen. Electromyogram (E.M.G.) including palate (Dr. D. Y. Mackenzie) normal. Muscle biopsy (Dr. M. Bodian): " no evidence of progressive degenerative process ; appearances suggest a congenital hypoplasia." Child discharged from hospital and made slow progress. Tube-fed until 4 years. Never able to walk unaided, but at 4 years 5 months would walk along furniture and ride a tricycle. Speech: spoke words at 18 months, but diction unclear. Readmitted to hospital at 4 years 5 months, when facial and palatal paresis again noted. Developed respiratory infection in hospital and died at 4 years 6 months. Necropsy refused.

Case 2.—Girl. Normal pregnancy and delivery. Birth weight 7 lb. 8 oz. (3,400 g.). Difficulty in swallowing, with regurgitation of fluids from birth: referred Hospital for Sick Children with unconfirmed diagnosis tracheo-oesophageal fistula at age 3 days. Palatal palsy noted. Required tube-feeding for some weeks. Floppy from birth. Did not walk unaided until 3 years 8 months. Speech developed normally but nasal in quality. On examination at 4 years 2 months: intelligent child, unable to hold up head properly ; marked immobility of face ; nasal speech ; mild palatal paresis ; gross generalized hypotonia ; all reflexes absent except ankle-jerks. E.M.G. (Dr. D. Y. Mackenzie) and muscle biopsy (Dr. M. Bodian) normal. I.Q. (Stanford-Binet) 128. When last seen at 5 years, skills gradually improving—able to ride a tricycle.

Case 3.—Boy. Normal pregnancy. Short (one hour) labour. Birth weight 8 lb. 4 oz. (3,740 g.). In good condition at birth.



Case 3. Showing wasting of tongue and strabismus.

Left talipes equinovarus. Sucked poorly and had restricted facial movement. Weaning difficult as unable to chew and swallow. Walked unaided at 3 years. Slow to learn to speak, and speech indistinct. On examination at 5 years 6 months: bilateral external ophthalmoplegia ; complete loss of facial expression ; gag reflex absent—palate did not elevate normally ; speech indistinct and unable to chew normally ; marked atrophy of tongue (see Fig.) ; weakness and wasting of left leg, with left pes cavus and talipes equinovarus ; no generalized hypotonia or other neurological signs.

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E.M.G. normal, no evidence of denervation. I.Q. (Merrill-Palmer) 95. Received speech therapy with slow improvement. No deterioration when seen last at 8 years 3 months.

Case 4.—Boy. Normal pregnancy and delivery. Birth weight 6 lb. 13 oz. (3,090 g.). As infant, regurgitated fluids through his nose. Not tube-fed. Feeding gradually improved, but at 4 years fluid still regurgitated through nose on occasion. Walked unaided at 20 months. Referred at 4 years because of absence of recognizable speech. Could not blow or suck through a straw. On examination: seemed intelligent, but no speech ; made noises with slightly nasal weak voice ; throat and palate moved, but not well ; slight weakness of upper and lower facial muscles ; no generalized hypotonia or other neurological signs. I.Q. at 7 years 6 months—non-verbal intelligence at least at educationally subnormal (E.S.N.) level. Given regular speech therapy, and attended E.S.N. school. At 7 years 11 months speaking very simple words, but still communicating mainly with gestures.

Case 5.—Boy. Normal pregnancy and delivery. Birth weight 7 lb. 12 oz. (3,515 g.). Born "full of mucus." Sucked well, but regurgitated fluids through his nose. Not tube-fed but continued to regurgitate until 4 years old. At 9 months given exercises to make him sit up properly, as head lolled. Walked unaided at 18 months. Tried to talk at 1 year, but at 3 years 6 months speech incomprehensible owing to marked nasal twang. On examination: rather dead-pan face ; marked nasal dysarthria although palate moved fairly well ; no generalized hypotonia or other neurological signs. Non-verbal I.Q. at 4 years 102. Received speech therapy. At 8 years 10 months nasality still marked, but some improvement.

Case 6.—Boy. Normal pregnancy and delivery. Birth weight 8 lb. (3,630 g.). No feeding difficulty reported at birth. Walked unaided at 19 months. Speech began between ages of 2 and 3 years. Speech therapy with slow progress since age 5 years. Referred at 7 years 9 months because of unintelligibility of speech. On examination: seemed of normal intelligence ; speech very indistinct largely because of nasal quality from palatal paresis ; could blow and whistle, but air escaped down nose ; facial muscles normal ; no generalized hypotonia or other neurological signs.

Case 7.—Girl. Normal pregnancy and delivery. Birth weight 7 lb. 2 oz. (3,230 g.). After birth mucus welled out of mouth and she had feeding difficulty. Tube-fed for 4 months. Several attacks of pneumonia in first few months of life. By 9 months chewing and swallowing normally. Walked unaided at 15 months. Spoke six words at 18 months, but speech always indistinct. On examination at 4 years 1 month: slight micrognathia with abnormal bite ; palatal paresis marked; no facial weakness ; no generalized hypotonia or other neurological signs.

Case 8.—Boy. Normal pregnancy and delivery. Birth weight 6 lb. 12 oz. (3,060 g.). Sucked well, but nasal regurgitation on feeding persisted until 9 months. Walked unaided at 16 months. Spoke six words at 2 years and sentences by 3, but referred at 5 years 10 months for poor speech, unintelligible to strangers. Could not blow a musical instrument unless he pinched his nostrils. On examination at 5 years 10 months: face lacked expression ; nasal voice ; palatal movements feeble ; air escaped down nose when he blew a pipe ; no generalized hypotonia or other neurological signs. Binet I.Q. 90. At 6 years making good adjustment at normal school. Receiving speech therapy.

Case 9.—Boy. Normal pregnancy. Prolonged (72 hours) labour. Birth weight 6 lb. (2,720 g.). At 6 weeks sucking noted to be poor, and was spoon-fed. Choking attacks due to accumulation of mucus in throat. Dribbling from mouth began at 4 months. Walked unaided at 18 months. Spoke single words at 2 years, but always very indistinct. Speech therapy began at 3 years 6 months. On examination at 7 years 7 months: an intelligent boy ; micrognathia ; nasal intonation of speech with poor articulation of consonants ; very little facial movement; soft palate hardly moved; tongue small and wasted ; no generalized hypotonia or other neurological signs. His speech improved, but remained nasal. When last seen at age 14 years 9 months was doing well at grammar school.

Case 10.—Girl. Normal pregnancy and delivery. Birth weight 6 lb. 9 oz. (2,975 g.). Regurgitation of milk down nose from birth. Excessive mucus production and poor sucking. Feeding difficulty disappeared by 9 months. Walked unaided at 2 years 3 months. Referred at 5 years 10 months because "slow in uptake" and speech defect. Understood perfectly, but speech unclear, especially consonants. On examination: voice had nasal twang ; blew whistle, but air escaped down nose ; palate elevated quite well ; no generalized hypotonia or other neurological signs. Hearing normal. I.Q. (part Stanford-Binet, part Wechsler) 80. Received speech therapy with gradual improvement.

Discussion

Feeding.—The feeding difficulties at birth are produced mainly by palatal palsy, although the occasional presence of micrognathia and the frequent association with facial weakness (discussed below) make the mechanics of the problem more complex. The prognosis for this aspect of the problem appears uniformly good, although tube-feeding may have to be persisted in for months or even years.

Speech.—The speech of these children has been likened by those who have heard it to that occurring with cleft palate, and, of course, failure to separate off nasopharynx from oropharynx produces the dysarthria in both conditions. Consonants are most affected, but nasal vowels such as oo (u) and ee (i) are often distorted. Here also the prognosis is slow recovery. In the absence of untreated controls it is impossible to say how much specialized speech therapy is able to contribute. There is no doubt that patients and their parents feel much benefit from encouragement given by skilled therapists. Pharyngoplasty has been advocated in similar cases, but, although the possibility of the operation being performed was discussed in many of these children, none underwent it, and I cannot therefore make any comment about its efficacy on the basis of this study.

Intelligence.—This was often called in question, as the facial immobility of some of these children made them look stupid. In fact, intelligence was present in a distribution similar to that of the general population. Indeed, rather satisfactorily, the average I.Q. of the four children actually tested was 100.

Associated Abnormalities.—Facial immobility, affecting the upper and lower parts of the face in both voluntary and involuntary movements was present in seven of these cases. Its severity ranged from total facial paralysis without a trace of movement to a strong suspicion that the "dead-pan" look of the child was outside the range of normality. In those cases followed up for any length of time the mobility of the facial muscles appeared to improve. In two of these children there was, in addition to Möbius's syndrome, a considerable degree of generalized muscular hypotonia. In both of them muscle tone improved as time went on, but one child remains seriously disabled, and the other died of a chest infection, a complication to which all these children are susceptible. There was no progression of the neurological illness in this latter case, although the fatal termination was certainly precipitated by the neurological disability. Micrognathia was present in two children and talipes equinovarus in two. These abnormalities have been reported in association with Möbius's syndrome.

Aetiology

The decision whether to label these cases a variety of Möbius's syndrome (facial diplegia) is a semantic problem. Möbius and others, later describing the same syndrome, regarded facial paresis as the primary feature in that condition, whereas in the present series bulbar symptomatology is the salient disability, facial paresis being a common accompaniment.

In the above cases where electromyographic studies were carried out there was no evidence of a denervating process. This disorder is probably a myopathy of congenital and non-progressive type. Evans (1955), discussing the aetiology of Möbius's syndrome in an article paradoxically entitled "Nuclear Agenesis," discards the possibility of primary nuclear involvement and persuasively proposes the concept of faulty development of branchial-arch musculature occurring at a critical phase in the growth of the embryo. The presence of severe universal

muscular hypotonia in two of these cases is notable. It argues against Evans's hypothesis, and prompted the suggestion put forward by Dr. P. H. Sandifer that a unitary concept is required to explain these three separately described conditions—"benign congenital hypotonia" or amyotonia congenita, Möbius's syndrome, and flaccid bulbar palsy of muscular origin. This suggestion is supported by the evidence presented here, for not only do these 10 children commonly show facial paresis, but their motor development is significantly retarded even where hypotonia is not flagrant enough to excite comment.

None of these children walked unaided before the age of 15 months, and the average age at which this skill was achieved was 2 years 3 months. Excluding those cases where hypotonia was detectable the average age was 21 months. In the absence of any other explanation—for example, cerebral palsy, mental subnormality—it seems possible that the skeletal musculature shares or shared the same defect as the bulbar and facial musculature. With lessons learnt from recent advances in our understanding of the progressive muscular dystrophies, one looks most hopefully to histochemical methods to elucidate the nature of this defect. The fact that this hypothetical defect reveals itself in so many different sites need not encourage speculation about possible multiple pathology, for the same capriciousness can be seen in other maladies (such as myasthenia gravis) which are recognized to be caused by single pathology.

Summary

The cases of 10 children with congenital, non-progressive, flaccid bulbar palsy, at first producing feeding difficulties with regurgitation of fluids, and later disturbances of articulation, are described. The prognosis of the condition is benign if respiratory complications do not occur, but the child may be left with a permanent dysarthria. The children have normal intelligence, but their gross motor development (as assessed by the age at which they first walked unaided) is considerably retarded. The condition was associated with facial diplegia (Möbius's syndrome) in seven of the cases, and with benign congenital hypotonia in two of them. The presence of a single defect responsible for all three associated conditions is postulated.

I should like to express my gratitude to Dr. P. H. Sandifer for the help and encouragement he has given me in the writing of this paper. Sir Wilfrid Sheldon kindly gave permission to publish details of the first case, which was under his care.

References

Benson, P. F. (1962). *Proc. roy. Soc. Med.*, **55**, 237.
Evans, P. R. (1955). *Arch. Dis. Childh.*, **30**, 237.
Henderson, J. L. (1939). *Brain*, **62**, 381.
Ingram, T. T. S. (1959). *Arch. Dis. Childh.*, **34**, 444.
Möbius, P. J. (1892). *Münch. med. Wschr.*, **39**, 17.

Clinical and Therapeutic Aspects of Kerosene Poisoning: A Series of 200 Cases

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Ingestion of kerosene is the commonest form of accidental poisoning encountered in African patients in Bulawayo. Of 42 cases of accidental poisoning admitted to Mpilo Central Hospital during the first nine months of 1963, 32 were due to the effects of kerosene ingestion—an incidence of approximately 75%. This trend had been apparent in previous years, and from August 1958 until August 1963 200 cases of kerosene ingestion have been admitted. This condition has been reported to occur frequently in other centres in Southern Africa (Kossick, 1961; McDonald, 1961). Kerosene is widely used for lighting, cooking, and heating purposes in the African townships of Bulawayo. Ingestion usually occurs in young children as a result of the ready availability of kerosene stored in various household utensils, such as cups, tins, and bottles.

The main controversy in the management of these cases is related to the place of gastric lavage in treatment (*B.M.J.*, 1963). The present study has been undertaken to assist in the assessment of this aspect.

Present Investigation

The 200 cases of kerosene poisoning admitted over a period of five years form the subject of this investigation.

Age Distribution.—The average age was 19 months, the majority falling within the range of 5 months to 5 years. The series included three adults, aged 20, 27, and 40 years, who had drunk relatively large quantities of kerosene in attempts at suicide.

The monthly incidence of kerosene poisoning showed a definite seasonal variation over the five-year period. The numbers were uniformly high during summer (September to April), but there was a sharp decline during the winter months (May to August).

Clinical Features

Although histories were not reliable, particularly with regard to the quantity consumed, a story of ingestion of kerosene was usually obtained from the parents.

The signs and symptoms could be divided into three distinct categories:

1. *Constitutional Disturbance.*—Moderate pyrexia (99–103° F. (37.2–39.4° C.)) occurred in 95 cases, usually within 24 hours of ingestion. In some cases this was the only abnormality noted, without any clinical or radiological evidence of involvement of a specific system. The duration of pyrexia varied considerably, and bore no apparent relation to the severity of the illness. An illustrative case was that of a child aged 2½ years, who had a temperature of 104.5° F. (40.3° C.) on admission, and who was discharged well after 48 hours. There was no evidence of pulmonary involvement. Drowsiness was a prominent feature in a number of cases.

2. *Gastro-intestinal Involvement.*—A story of vomiting was frequently obtained, but the exact incidence could not be ascertained, owing to the inaccuracy of the history in many cases. Diarrhoea was uncommon, occurring in only six cases.

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