Bilateral Congenital Palsy of the Abducens and Facial Nerves

A Case Report*

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Attention has been focused on congenital nuclear palsies for eighty years, but we do not yet have any satisfactory explanation of their pathogenesis and etiology. Recently we observed a case of congenital abducens and facialis palsy.

S. B., a girl three and a half years of age, was admitted to the Hacettepe Children's Hospital (Case No. 57-1418) on October 2, 1957. According to her parents' statements, she was not normal even at birth, as a seven-month premature. Delivery was normal. The birth weight was not known. She could not open her eyes for two months and after opening gradually they have never closed nor blinked since. The eyelashes turned inward. She sucked with great difficulty. The eyeballs deviated upward. When crying or laughing her expression did not change and there was no lacrimation. She could not masticate easily. Her facial expression had not changed since birth.

Family history: The father, aged 24, is in good health, as is the mother, aged 23. The patient has one sister aged 11 months and in good health. The father is a seller of "yoğurt" (a dairy product), and the family's financial situation is rather poor.

Physical examination: Weight 9.6 kg, height 86 cm. Temperature 38 degrees Centigrade. The patient was moderately well developed. Her cheeks were flabby and swollen; the nose continued uninterruptedly with the forehead and its tip was flat. The mouth was always open and the upper lip protruded, so that the whole appearance of the mouth was that of a triangle. The lower teeth were in poor condition. The uvula and the ears appeared normal.

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The cranial nerves were systematically examined. No olfactory tests could be performed but she was sensitive to irritants. Vision appeared normal. An ophthalmologist examined the third, fourth and sixth nerves and reported: "Trichosis on the nasal portion of the right upper lid, moderate shortness of both lower lids, strabismus on upward gaze. There is no lateral movement of the eyeballs and no blinking reflex. Opalescent patches on the corneae are present due to failure of lacrimation and inability to close the eyes." Strabismus was more prominent in the right eye. Vertical movement of the eyeballs was limited but present, and was performed more easily during associated movements. Study of the seventh nerve revealed no movement nor expression of the face. The eyelids were always open, even during sleep. When she attempted to close her eyes the eyeballs deviated upwards. There were very slight downward movements at the corners of the mouth. The other cranial nerves showed no significant findings save that mastication and lateral movements of the chin were rather weak. There were no disorders of sensation. The deep reflexes were diminished.

The liver extended about one inch below the costal margin. As noted in the photograph, pigmentation areas were present on the the abdomen.

X-ray examinations, blood counts, the blood Wassermann reaction and the urinalysis were all normal. The cerebrospinal fluid showed 66.6 mgm% protein but other tests, including the Wassermann reaction, were normal.

Her speech was unintelligible due to the lack of labial sounds. Electrodiagnostic tests could not be performed because of the child's excitement.

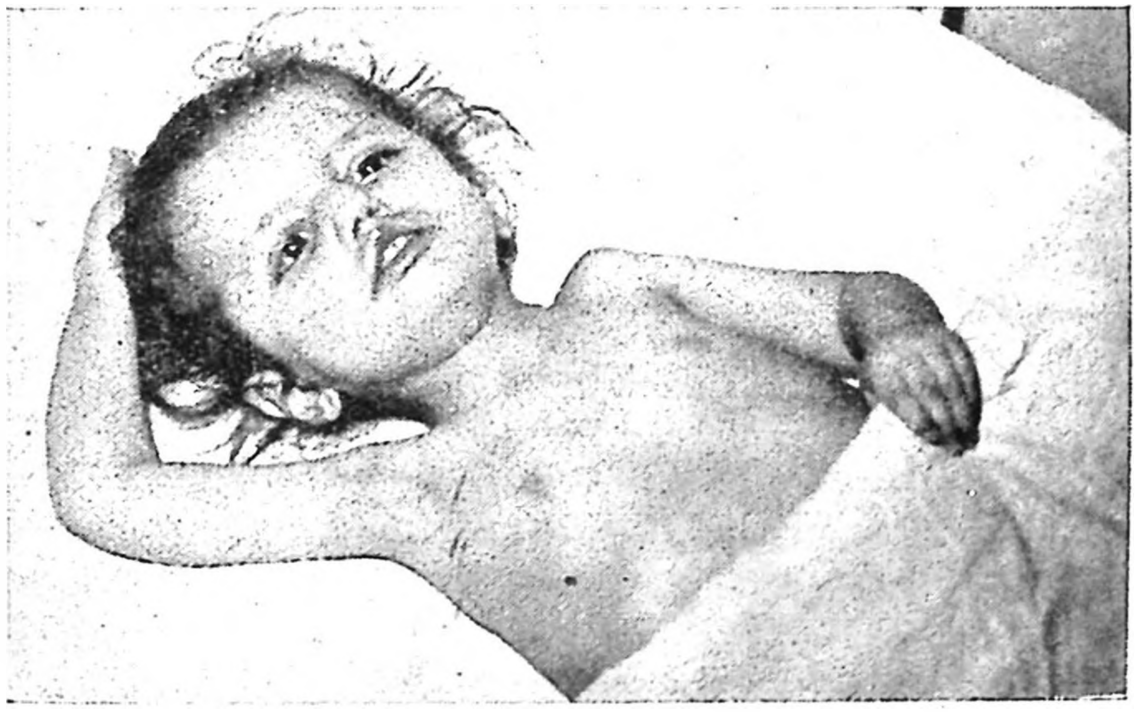
In summary, this girl of 3.5 years has a congenital complete double palsy of the sixth and seventh cranial nerves with lack of lacrimation, and paresis of the third and fifth cranial nerves. The birth was normal save for prematurity. Her infancy was almost normal save for the above disorders. Because her symptoms were present at birth and are bilateral, laboratory findings are normal, and save for isolated cranial palsies the neurological examination is negative, we diagnosed her case as one of congenital nuclear palsy due to aplasia.

Nuclear palsies show themselves in various clinical types. Moebius [2,3] has classified them, and Henderson [1] in 1939 collected 60 cases from the literature and added one himself.

Nuclear palsies are sometimes associated with body and extremity deformities. They are seldom complete and symmetric. Lack

of lacrimation, as shown in our case, is rare. Henderson cited three such cases and Robertson [4] mentioned one.

The pathogenesis is very difficult to explain by either infectious or hemorrhagic proce sses,neither of which are so selective and localized. There are other possible explanations (toxic, aplastic, or excess secretion of the choroid plexuses) but none of these is very satisfactory.



REFERENCES

1. Henderson, J. L.: The Congenital facial diplagia syndrome: Clinical features, pathology and etiology. A review of sixty-one cases, Brain 62: 381-403, 1939.
2. Moebius, P. J.: Über angeborene doppelseitige Abducens-Facialis Lahmung, Münchner Medizinischer Wochenchrift 6: 91-94, Feb. 7, 1888, ibid, 7: 108-111, Feb. 14, 1888.
3. Moebius, P. J.: Über infantilen Kernschwund, Münchner Medizinischer Wochenschrift 2: 18-12, Jan. 12, 1892; ibid, 3, 41-43, Jan. 19: 1892; ibid, 4: 55-58 Jan. 26, 1892.
4. Robertson, S. E. J.: Congenital facial diplegia (Moebius' syndrome), Med. J. Australia 2: 335-337, Aug. 29, 1953.