HEMIPLEGIA IN CYANOTIC CONGENITAL HEART DISEASE

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Although the cases to be described lack any anatomical observations, the association of uncomplicated cyanotic congenital heart disease with the sudden onset of hemiplegia appears worthy of record. All three cases have only recently come under our observation.

NOTES OF CASES

Case 1.—J. L. was first seen at the age of eight months. He was a first child, and because of the mother's "kidney trouble" the pregnancy was terminated three weeks early. Shortly after birth it was noticed that the hands and feet went blue at times and that when crying he appeared to get unduly out of breath. He had gained weight satisfactorily. On examination there was no marked cyanosis and the only clinical abnormality was a loud central systolic murmur. On X-ray, there was a heart shadow suggesting the tetralogy of Fallot. A month later he was admitted to Southmead Hospital because, following a particularly intense attack of cyanosis and dyspnoea five days before, it was noticed that the right arm and leg did not move normally and that on crying the face was drawn over to the left side. On admission there was a right hemiplegia. The cardiac physical signs were unchanged, there was no fever, and no other abnormality was found. He continued to have attacks of cyanosis of varying severity. A lumbar puncture a week after admission was blood-stained—apparently traumatic—but otherwise normal. A blood count showed 6,000,000 red cells with a haemoglobin of 90 per cent. Since then there has been no change.

Case 2.—D. B. was born normally and was found to have congenital heart disease at the age of three months. He progressed favourably except for attacks of cyanosis until the age of eleven months, when loss of power in the right arm and leg was noticed a week after an attack of bronchitis. He was admitted to hospital the next day and found to have a right hemiplegia. The cerebrospinal fluid two days after the onset showed 6 lymphocytes per c.mm. and 50 mg. of protein per 100 c.c. Ten days later this was normal. There

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was a mild pyrexia for three weeks. Since then the child has remained the same. There is constant cyanosis with exacerbation in attacks. There is dextrocardia with a basal systolic murmur. X-ray examination shows a boot shaped heart with dextrocardia. Electrocardiogram shows right axis deviation when allowance is made for the dextrocardia. A recent blood count shows 9,000,000 red cells with a haemoglobin of about 160 per cent. This is probably a case of the tetralogy of Fallot with dextrocardia.

Case 3.—M. W. was first seen at the age of nine years. He is deeply cyanosed with a soft basal murmur. X-rays show a rather boot shaped heart with a small pulmonary arc. His electrocardiogram shows marked right axis deviation. He has 7,000,000 red cells per c.mm. and a haemoglobin of 140 per cent. The X-ray picture suggests that this may be a case of Fallot's tetralogy but the murmur is so soft as to raise doubts about this diagnosis. When first seen he had a right hemiplegia which was at first regarded as congenital. However, on further enquiry his mother states that this was first noticed at the age of eight months, after a week's illness in which he "seemed dazed."

Discussion

This syndrome was entirely new to us, and the only references found in the literature are the cases reported by Ford (1937) and the case recently reported by Wood (1942).

Ford's first case was a mentally defective child dying at the age of two and a half of pneumonia. He exhibited athetoid movements at the age of twelve months and the colour is said to have been "dusky." Post-mortem, there was a patent ductus arteriosus and a patent interventricular septum. The left middle cerebral artery showed an old thrombosis, partly recanalized, with an infarct involving the motor cortex and part of the temporal lobe and extending into the internal capsule and lenticular nucleus.

His other case was a child, blue from birth, who at ten months showed a basal systolic thrill and murmur and a red cell count of 7·5 millions. At two years of age he suddenly became stuporous and developed a left hemiplegia. A few days later he became more comatose and showed a bilateral extensor plantar response. He died shortly after. Post-mortem, there was atresia of the pulmonary artery, patent interventricular septum patent foramen ovale, and cardiac hypertrophy. There was a thrombosis of the right middle cerebral artery with infarction, and a later thrombosis of the superior longitudinal sinus and a small infarct involving part of the left motor cortex.

Wood's patient had been cyanosed from birth. There was no definite history of the onset of hemiplegia, but this apparently developed in infancy, since she had never walked and did not sit up until two years of age. Post-mortem, there was no gross vascular lesion but degenerative changes apparently due to the intense congestion. Bodechtel (1932) has described changes in the brain in congenital and acquired heart disease, apparently due to anoxaemia, but these changes would hardly appear sufficient to produce hemiplegia.

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Ford attributes these thromboses to the polycythaemia and compares this with the cerebral thrombosis that may complicate polycythaemia vera in the adult. However, in our first case the polycythaemia was slight, and in Ford's first case the cardiac malformation should have produced little cyanosis except with the terminal pneumonia.

Since Wood's case showed no actual occlusion it appears that the brain lesion associated with cyanotic congenital heart disease may be produced in at least two ways. Obviously the possibility of embolism must be considered, but it would appear likely that if this were the cause, emboli elsewhere would have been observed. The possibility of a superimposed bacterial endocarditis seems to be excluded by the subsequent history. The actual pathology of the cases here reported must remain uncertain.

SUMMARY

Three cases of cyanotic congenital heart disease developing sudden hemiplegia in infancy have been described. The possible mechanism of this has been discussed.

We are indebted to Dr. R. M. Norman for drawing our attention to the observations of Bodechtel.

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