AGENESIS OF THE CORPUS CALLOSUM.

By J. S. Lloyd, M.D., M.R.C.P., D.P.M.,
Medical Superintendent, Fountain Hospital (L.C.C.); and

J. N. Jacobson, M.R.C.S., L.R.C.P., D.P.M., D.M.R.E.,
Deputy Medical Superintendent, Fountain Hospital (L.C.C.).

Agenesis of the corpus callosum is not as rare as might be expected and over a hundred cases are on record. It is, however, remarkable for the diversity of the clinical signs that are associated with it, and no doubt these are dependent on other anomalies of the brain, some of which apparently always accompany this condition.

Case-Report.

The patient was a female idiot who died at the age of nine years of pneumococcal meningitis. She was microcephalic, the cranial circumference being 17 in., and in addition she had the configuration of the skull and loose scalp of microcephaly. She had spastic quadriplegia with the usual signs of upper motor neurone lesion, and double optic atrophy with absent light reflex. Several degenerative phenomena were in evidence, including facial asymmetry, strabismus, scoliosis and attacks of *grand mal*. Post mortem, a large fibroma was found in the wall of the left ventricle of the heart which seemed almost to obliterate the cavity, but had given rise to no murmurs or other cardiac signs during life.

Description of the Brain.

Pneumococcal meningitis was evidenced by hyperæmia and clouding of the meninges with purulent exudate over the convexity of the brain, from which pneumococci were cultured. The cerebral hemispheres were small, weighing only $8\frac{1}{4}$ oz. each. The convolutions on the external surface of the cerebrum showed no marked abnormality, and in particular the pre-Rolandic area seemed well developed. Microgyria was absent. The internal surface showed very well the radial arrangement of sulci and non-union of the parieto-occipital and calcarine fissures which are characteristic of the condition. On separating

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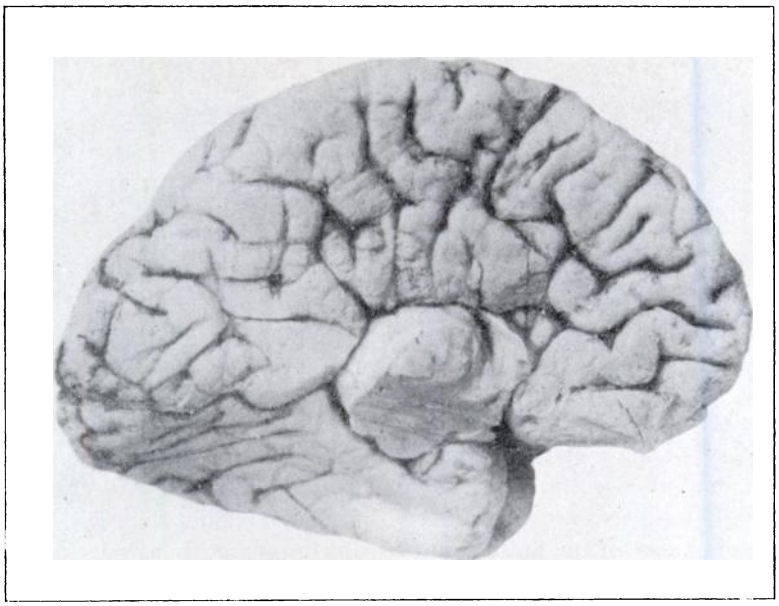

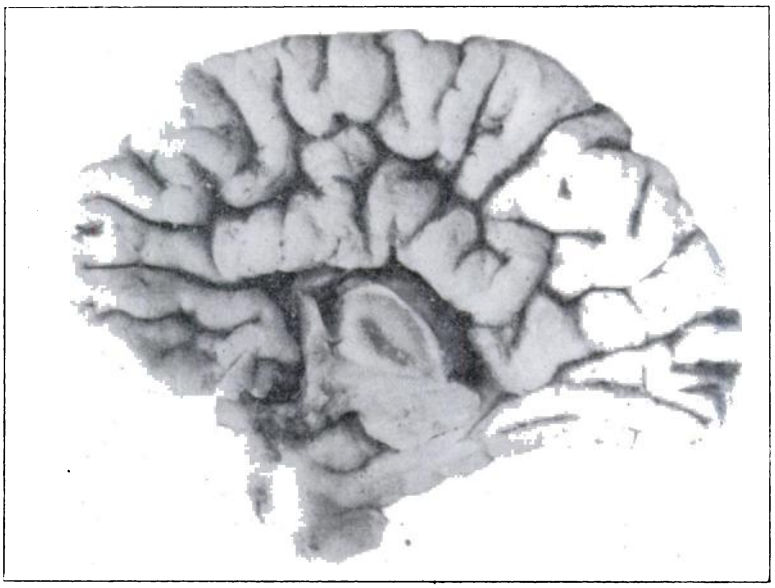

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the hemispheres, complete absence of the corpus callosum was revealed. The fornix was present but small, and the body appeared to be in two separate halves. A rudimentary septum pellucidum was attached to the anterior aspect of each half of the body. The anterior commissure was not enlarged. The posterior horn of the lateral ventricle was dilated, but the internal hydrocephalus was not pronounced. The olfactory tracts were present, although very slender, and the origins of the other cranial nerves showed no abnormality. The cerebellum was well developed.

Discussion.

No clue was obtained as to the ætiology of the condition. The family history was unsatisfactory. The only sibling was stillborn in association with placenta prævia.

Agenesis of the corpus callosum may be partial or complete, and the recorded cases are divided about equally between these groups. The expectation of life is limited to childhood, although some cases have died at an advanced age.

Various grades of intelligence have been found with both partial and complete agenesis and without correspondence to the degree of agenesis. Tumour of the corpus callosum is known to be associated with mental disorder in the nature of dementia. The present case throws no light on this point: the patient was a profound idiot with a mental age of less than six months, but this may well have been due to any of the other anomalies of the brain which were present. It is usually believed that apraxia results from disorder of the corpus callosum, but apraxia could not, of course, be assessed in this case. The spastic quadriplegia that was present is in accord with the result of section of the corpus callosum in experimental animals but is at variance with the usual symptoms in agenesis. Good physical development seems to be the rule, although some catastrophe usually completes the case in the first decade. Anatomically these cases have several very constant features, some of which are well shown by the brain illustrated. On the internal surface of the cerebrum the radial arrangement of the fissures and the failure to unite of the calcarine and parieto-occipital fissures have usually been noted, with either partial or complete agenesis. Microgyria and anomalous pattern of the cortex on the external surface of the cerebrum, absent in this case, are usually present. The fornix seems always to be present and is frequently normal in appearance, and the anterior commissure is frequently enlarged. Neither of these conditions obtained in this specimen, and the cerebral hemispheres were practically ununited. The specimen also did not show the absence of the olfactory bulbs, which is a common feature, but showed the usual enlargement of the posterior horn of the lateral ventricle.

It is interesting to note that ante-mortem the patient presented the ordinary signs of microcephaly, but the small size of the brain was presumably due to the absence of the corpus callosum and its connections.

SUMMARY.

1. A case of complete agenesis of the corpus callosum in a quadriplegic idiot aged nine is recorded.

2. The brain is described and a resumé given of previous anatomical and clinical findings in the condition.

3. The association of a tumour of the heart and various stigmata of degeneration is noted.

The following are references in English to agenesis of the corpus callosum:

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