ANENCEPHALY IN IDENTICAL TWINS

BY J. E. JOSEPHSON,

Department of Physiology, Queen's University, Kingston, Ont.,

AND K. B. WALLER, M.D.,

Rockwood, Ont.

ANENCEPHALY occurs very rarely in twins. Occasionally case reports describe twins of which one is anencephalic, as in the case by Thompson.[1] Less frequently conjoint twin anencephalic monsters are described. Mudaliar[2] describes one such, a thoracopagus dibrachius dipus, an anencephalic female monster, fused from the root of the neck downwards, with separate heads. A careful search of the literature has, however, revealed no previous description of separate twins, both anencephalic. The twins described herewith were delivered by one of us (K.B.W.).

CASE REPORT

Family history.—The father is a laborer. The mother, now 31 years of age, is one of thirteen children. One of her brothers has a club-foot. She has been confined twelve times, with two sets of twins. She has never had a miscarriage or abortion. There are twelve living children. The first-born are twin boys, 15 years of age, who had a criminal record and were sentenced to Mimico Industrial School. Since then they have been transferred to the Ontario Hospital for Feeble Minded at Orillia. The next child is a girl who is unable to speak plainly, the next a boy who is also unable to articulate clearly. He is very small of stature, but quite strong and exceedingly pugnacious. He has been caught stealing various times. All the children exhibited an impediment in speech on reaching the talking age, and are backward in school. Since the anencephalic twins were born the mother has been delivered of one child in good physical condition.

The anencephalic twins were born (the ninth confinement), when the mother was twenty-nine years of age. So far as she knew they were full term, but she has menstruated so rarely in her married life that she has had difficulty in calculating the date of conception of her children. The physician was not called until she had been in labour one-half hour. A marked hydramnios was recognized, and she was conveyed to hospital. Twin presentations were diagnosed, one vertex, R.O.P. and one face, L.M.A. Forceps were applied in both cases, but failed to hold, and the twins were born spontaneously. There was only one placenta, the babies were both females, and probably identical twins.

EXTERNAL FEATURES OF THE TWINS

For convenience we shall designate the twins "A" and "B".

Twin "A".—The weight of twin "A" was 940 grm. This twin is typical of iniencephaly. Because of the extreme dorsiflexion of the head on the cervical and thoracic vertebræ, the nose is the highest point. The lower thoracic and lumbar vertebræ show marked kyphosis, thereby giving the whole spine a sort of S-shape.

There is an extreme spina bifida, extending the length of the cervical region and including most of the thoracic. Overlying this is a large and delicate fleshy mass, measuring 63 by 55 by 22 mm. which protrudes from a deficiency in the cranium which should be occupied by the occipital bone. Two membranes, the outer tough, the inner delicate, surround the herniated mass, and blend with the lining of the cranial cavity. Microscopic section of the mass shows it to be angiomatous, consisting for the main part of blood sinuses and vessels filled with clot, while an occasional degenerated neuroglia cell is found here and there.

The shoulders are carried high, and, there being no neck, the head rests entirely on them. There is a good growth of hair on the scalp and an abundance of subcutaneous fat, especially between the face and thorax, giving the fetus the appearance of having a double chin. In the "anencephalic syndrome", described by Browne,[3] characteristic features are gaping of the mouth, with protrusion of the tongue and bulging of the eyes. In this case however, while the mouth is gaping, there is no protrusion of the tongue, and while there is considerable thickening of the eyelids, there is no actual bulging of the eyes. The frontal bone, instead of rising perpendicularly, is rather flattened backwards in the vertical plane at an angle of about 15 degress to the orbital plate.

Twin "B".—Twin "B" weighed 875 grm. This twin is typical of the microcephalus acranius described by Nanagas.[4] There is no protruding hanging mass, which, together with the fact that it is less obese than "A", probably accounts for the difference in weight. What obesity there is is noted mainly in the upper thoracic and cervical regions. The head, instead of being dorsiflexed as in the case of the other

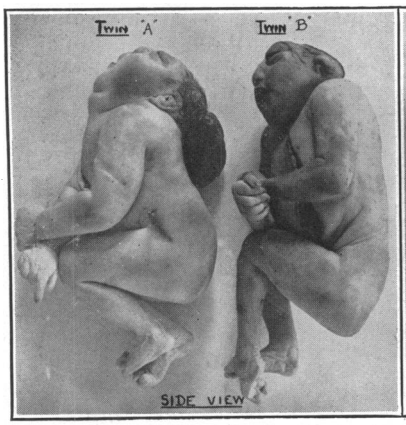

FIG. 1.—Side View.

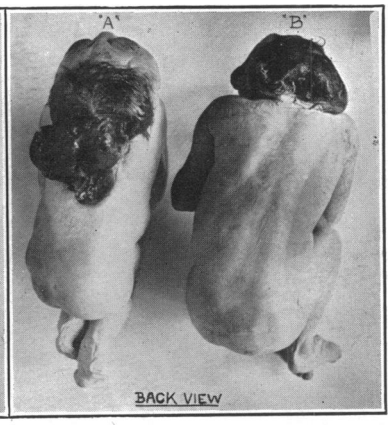

FIG. 2.—Back View.

twin, sits upright on the shoulders. There is no neck, but a deep groove running ventrally from ear to ear separates the head and thorax. The jaw sags and rests on the chest, but there is no protrusion of the tongue. The pinnæ of the ears are large and pendent, almost coming to rest on the shoulders. The eyes bulge and the nose is flattened. The shoulders are carried high, are slightly rounded, and covered with fine hair. The frontal bone, overlying which there is a considerable amount of subcutaneous fat, slopes backwards immediately above the orbits, giving the fetus the appearance of lacking a forehead. The top of the skull is completely deficient, the basis cranii being covered over by a thin, flattened, hard and fleshy mass, 47 by 54 mm., which in turn is covered over by membranes. The mass is surrounded by a deep groove anteriorly which becomes shallower laterally. There is no hair covering the mass, the growth of hair being confined to an area immediately outside the groove. Microscopically the mass resembles that of twin "A", *i.e.*, it mainly consists of blood sinuses and vessels. A small cyst showing some attempt at glandular formation was found embedded in the lower and posterior part of the mass. It weighed 35 mgrm. and measured 4 by 4 by 2.5 mm. The stubs of two cranial nerves were also found in the two small openings in the basis cranii under the mass.

A comparison of the skulls of the twins shows "A" to have a more complete skull than "B", although x-ray plates showed that the bones of the former were badly formed. The mass covering the basis cranii of "B" leaves no cranial vault at all, whereas the cranial vault of "A" is intact except for a deficiency in the occipital bone where the protruding mass issues. There is no spina bifida and the limbs show no malformations.

INTERNAL FEATURES AND ANOMALIES

Twin "A" shows a rare anomaly, in that there is a deficiency of the diaphragm on practically the whole of the left side, with only a thin span of muscle crossing from right to left on the anterior and lateral abdominal walls. This leaves a large opening through which the thoracic and abdominal cavities communicate. Through this opening the following abdominal structures protrude into the thoracic cavity:— (1) the whole left lobe of the liver; (2) the main part of the transverse colon and great omentum. The latter is frayed, ragged, and quite short, and does not come down as a fold over the intestines; (3) the spleen, located at the apex of left thoracic cavity and situated deep to the liver and transverse colon; (4) a large portion of the small bowel; (5) the stomach and first part of duodenum; (6) the pancreas. The normal thoracic viscera are pushed over towards the right half of the thorax. The heart is on the right and superior to it lies the thymus.

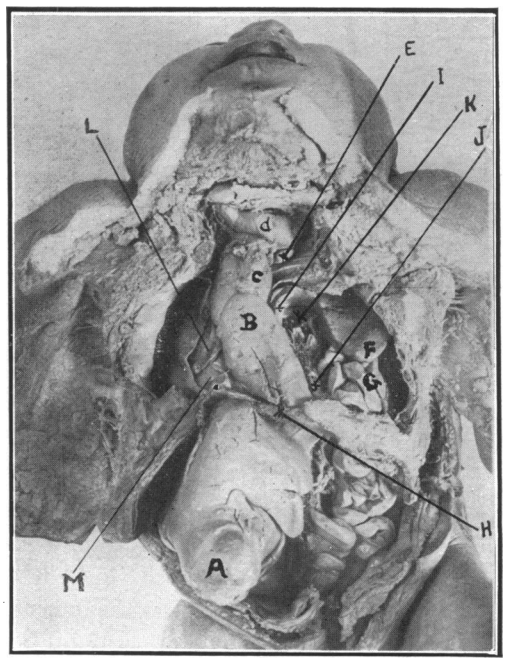

FIG. 3.—Dissection of twin "A" showing:— A. knob of liver herniating the umbilicus; B. left lobe of the liver; C. thymus; D. thyroid; E. accessory thyroid; F. spleen; G. small intestines; H. thin span of diaphragm; I. left lung; J. stomach and first part of the duodenum; K. transverse colon; L. right lung; M. heart.

The lungs are small and flattened, the left one being wedged in between the spleen, stomach, and left lobe of the liver on one side, and the heart and thymus on the other.

Ballantyne,[5] however, has shown that left-sided diaphragmatic defects are quite frequent in iniencephaly. Arey[6] says that "the persistence of a dorsal opening in the diaphragm, more commonly on the left side, finds its explanation in the imperfect development of the pleuro-peritoneal membrane."

Twin "A" also has an umbilical hernia containing a rounded knob of the right lobe of the liver, measuring 1.5 cm. in diameter.

Twin "B", on the other hand, shows very little deviation from the normal in regard to its gross internal anatomy, except for an exceptionally large left lobe of the liver, large, solid and congested-looking lungs, and a double right ureter. The thymus is made up of three lobes, but sections of it appear quite normal. Along the lower border of the thyroid gland there are four or five small bodies, the largest of which is about the size of an orange seed, and which on section proved to be an accessory thyroid. The others are small and were not examined microscopically.

The Adrenals.—Since the suprarenals and pituitary have been described by various authors as the endocrine glands showing the most change in anencephaly, a brief description of these glands will be given. The classical description of anencephaly always associates the condition with absence or poor development of the adrenal glands. Browne[3] says that they may be entirely absent on one or both sides. Vaclav[7] found the adrenals in 55 of 56 cases examined. Kiyono[8] found them present in 11 cases, whereas Ettinger and Miller[10] investigated 9 cases and in 2 they were entirely absent. When present, the chief disturbance lies in the cortex; the medulla is considered to be microscopically normal.

Landau[12] and Vaclav[7] describe the anencephalic adrenal cortex as being diminished in size, but precociously developed, in that there is a premature development of the permanent cortex, with a disappearance of the fetal boundary zone described by Elliott and Armour,[11] Landau[12] and Cooper.[13] Kiyono,[8] Ettinger and Miller,[9, 10] and others find the gland markedly underweight in all their cases. Bär and Jaffé[14] noted in 6 anencephalics of about the 7th to 8th fetal months, that lipoids were always present in large amounts.

In our case both suprarenals were found in each twin, always at the upper pole of the kidney. No accessory cortical nodules were found. The weight and measurements of each gland were as follows.

TWIN "A"

	Measurements	*Weight*
Left Adrenal	—19 by 8 by 5 mm.	0.355 grm.
Right Adrenal	—20 by 15 by 5 mm.	0.56 grm.

TWIN "B"

	Measurements	*Weight*
Left Adrenal	— 16 by 5 by 2 mm.	0.115 grm.
Right Adrenal	— 11 by 4 by 4 mm.	0.063 grm.

The normal weight of the adrenals at birth as given by Kiyono[8] is 2.5 to 3 grm. Microscopically, there were no essential differences in the arrangement or structure of the cells in the adrenals of the two twins, although lipoids were present in greater amounts in those of twin "A". There is precocity of development of the cortex in each twin, in that the definitive zona glomerulosa and zona fasciculata occupy about half of the total area of the shrunken cortex.

The pituitary gland.—In most of the cases examined only a pars anterior has been found. A few cases are reported in which the posterior lobe was noted. Browne[3] in his series of 5 cases found no trace of a pituitary body. Kohn[15] examined 11 cases, in which the anterior lobe was invariably present, the pars intermedia absent, and in only 3 was any pars nervosa discovered. Ettinger and Miller[9, 10] found an anterior lobe in 8 of their series of 9 cases and, of these, 3 only showed a pars nervosa.

In our case only the pars anterior was found in each twin. In twin "B" the cranial portion of the cranio-pharyngeal canal persisted. In twin "A" the measurements of the gland were 70 by 80 by 20 mm., and the weight was 80 mgrm. In twin "B" they were 89 by 80 by 30 mm., and 70 mgrm. Histologically, the gland was composed principally of blood sinuses or lacunæ. Between the blood sinuses columns of epithelial cells consisting chiefly of chromophobes and non-granular chromophyles, of which the former predominated, made up the glandular parenchyma.

CONCLUSION

A case of anencephaly in each of female twins, probably identical, is described. The occurrence of the condition in both babies, born

of a family with low mental development, supports the theory that the condition is an expression of inherited tendencies rather than an antenatal accident. In spite of the gross cranial defect common to both twins there are extreme differences in bodily development, leading one to believe that although the same basic cause might determine the anencephaly in both cases, the later physical development might be affected by environmental conditions, such as the intrauterine relationship, one to the other.

A survey of the literature reveals no previous example of anencephaly in identical twins.

We wish to acknowledge the assistance of Dr. W. D. Hay for his careful preparation of the photographs.

REFERENCES

1. THOMPSON, *Boston Med. & Surg. J.*, 1925, **193**: 1241.
2. MUDALIAR, *J. Obst. & Gyn. of Brit. Emp.*, 1930, **37**: 753.
3. BROWNE, *Edin. Med. J.*, 1920, **25**: 296.
4. NANAGAS, *Am. J. Anat.*, 1925-26, **35**: 455.
5. BALLANTYNE, Ante-natal Pathology and Hygiene, Vol. II. The Embryo, Wm. Wood, N.Y., 1904.
6. AREY, Developmental Anatomy, W. B. Saunders, Phila., 2nd ed., 1930, p. 212.
7. VACLAV, *Sbornik Lekarsky*, 1927, **28**: 399.
8. KIYONO, *Virch. Arch. f. path. Anat.*, 1925, **257**: 441.
9. ETTINGER AND MILLER, *J. Obst. & Gyn. of Brit. Emp.*, 1929, **36**: 552.
10. ETTINGER AND MILLER, *Trans. Roy. Soc. Can.*, 1926, Sect. V, p. 249.
11. ELLIOTT AND ARMOUR, *J. Path. & Bact.*, 1911, **15**: 481.
12. LANDAU, *Centralbl. f. allg. Path. u. path. Anat.*, 1913, **24**: 404.
13. COOPER, Histology of the more Important Endocrine Organs at Various Ages, Oxford Univ. Press, London, 1925.
14. BAR AND JAFFÉ, *Centralbl. f. allg. Pathol. u. Anat.*, 1924, **35**: 179.
15. KOHN, *Arch. f. micr. Anat. u. Entwg.*, 1924, **102**: 113.

ACUTE PHLEGMON OF THE STOMACH AND DUODENUM*

BY A. V. GREAVES, M.B. (TOR.), D.T.M. (LIV.),

Government Bacteriologist,

Hong Kong

PHLEGMONOUS inflammation of the stomach and duodenum is a pathological lesion of considerable rarity and is always deserving of report, more especially since the etiology still remains undiscovered.

According to Leith[1] the first description of phlegmonous gastritis was given by Varandaeus in 1620 in his "Tractatus de Morbis Ventriculi", while later mention is made of it by Borel, Bonet and Sand (1656-1701). These earlier references appear to have been concerned with the circumscribed variety of the disease, whereas the diffuse form was not definitely described until 1839 by Andral, and later, in 1861, by Cruveilhier. Greater interest in the condition in modern times has resulted in the recording of larger numbers of cases, so that Anderson[2] in 1922 was able to collect 220 cases from the literature. Four cases, also, were reported by Pritchard and McRoberts[3] in 1931.

The analogous affection of the bowel, phlegmonous enteritis, is of rarer occurrence still. Thus MacCallum,[4] in 1906, was only able to collect 7 cases from the literature, 5 only of which affected the small bowel. In an excellent summary of the literature Irwin and McDonald[5] quote in some detail 21 cases of involvement of the duodenum and jejunum, and refer to 3 further cases of Bohmansson's together with 2 personal cases, making a total of 26 cases in all. In a review of continental literature Bohmansson[6] analyses 68 cases but of these only 37 involved the duodenum and jejunum alone. The rarity of phlegmonous enteritis is thus evident.

The two cases referred to here occurred in the post-mortem service of Dr. K. H. Uttley, Medical Officer in Kowloon, to whom I am indebted for the opportunity of studying them.

CASE 1

The only information obtainable in this case was a history of abdominal pain, vomiting and diarrhœa for three days previous to death.

The body was that of a fairly well nourished Chinese male about 35 years old. The skin had a subicteric tint and showed post-mortem staining. Abdominal section showed the presence of general peritonitis, but there was no free pus in the cavity. Between the stomach and the duodenum there was a small amount of thick fibrino-purulent exudate. The general peritoneum, both parietal and visceral, had the dull, granular appearance characteristic of early peritonitis. The great omentum was partly drawn up into the epigastrium and its vessels were definitely injected. The duodenum and beginning of the jejunum were strikingly swollen and were obviously the seat of the main pathological process in the abdomen. The first part of the duodenum was a dusky shade, the second part only slightly so; the remainder of the duodenum and the first part of the jejunum were definitely congested and pink. The peritoneum in this area was much thickened, and the adjacent lymphatic glands

* From the Bacteriological Institute, Medical Department, Hong Kong, Hon. Dr. A. R. Wellington, Director of Medical and Sanitary Services.