congenital dilatation of the colon. One was operated upon and succumbed. The present one he did not have operated upon, as he thought he would try medical means, and these also had not been successful. Immediately the bowels were cleared out the child steadily became worse and died, apparently from sapræmia. The mucous membrane was intact. He had been asked how such cases should be treated, and he had to confess that he did not know, as his experience had been limited and his results unfortunate.

Case of Acrocephaly, with other Congenital Malformations.

By GEORGE CARPENTER, M.D.

CHARLES G., a 5 weeks old infant, whose family history I will subsequently detail, is a weird-looking specimen of the human race. When looked at from the front, the face and skull form an ace of diamonds-shaped figure, or perhaps rather that of two right-angled equilateral triangles placed base to base, the inferior angle of one forming the chin, the superior angle of the other the top of the cranium, and the lateral angles gaps in the skull where the brain is in close contact with the overlying soft parts. The eyes protrude frog-like, the eyeballs being kept in position merely by the lids, so that it is possible to readily dislocate the globes and permit the organs to hang suspended by their muscular and nerve attachments (fig. 1). The bridge of the nose is much depressed, so that the nostrils look forwards. The palate is very high and the arch narrow. Such is the appearance of the front of the child's head and face. When viewed from the side the face and forehead are seen to slope backwards in a straight line from the tip of the chin to the top of the skull (fig. 2). There is no prominence of the forehead at all. The occipital bone is thrust forward somewhat, with the result that the occipital and frontal bones unite and form the peak of the skull. The parietal bones do not take any part in the conformation of the cranial vault (fig. 3).

Reviewing the cranial bones more minutely, the following anatomical characteristics are observable: the frontal bone has a central ridge about 1 in. broad running from below upwards, and on each side of it there is a shallow sulcus (*see* fig. 1). The bone itself is flattened (*see* fig. 2). The orbital ridges can just be made out with the finger and they are complete. But the orbital cavities are evidently shallow and ill-formed as judged by the prominent and readily dislocated eyeballs. The supra-orbital notches are unusually prominent. The malar bones, in so far as

they form the orbits, are not very pronounced to the feel, and the zygomata are not readily made out, though present, owing to the bulging of the brain in the neighbourhood of the temporal fossæ (*see* fig. 1). The occipital bone is not only thrust forward, but it is much deformed. Where the occipital and the frontal bones meet there are two knobs of bone, one on each side of the mid-line, about 2 in. apart, like the rudimentary horns of a young calf (*see* fig. 2). At the summit of each can be felt a distinct suture. There are also some other irregular

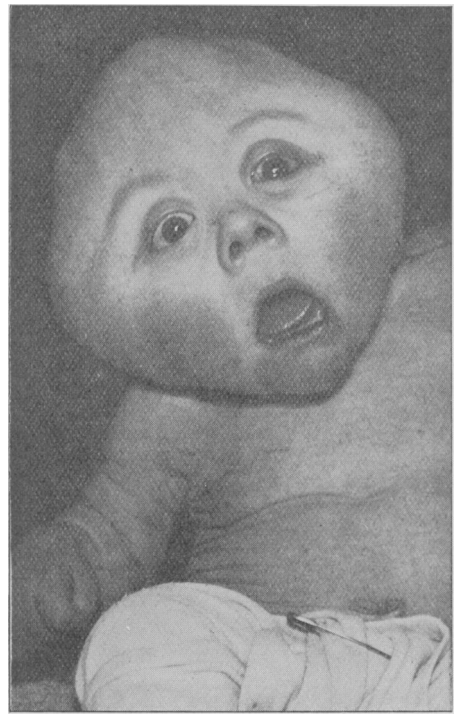

Fig. 1.

bony thickenings on each side along the occipito-parietal sutures (*see* fig. 2), and all along the fronto-parietal suture on each side is a definite raised ridge (*see* fig. 3). There is no anterior or posterior fontanelle. A large prominent mass of bone can be felt corresponding to the external occipital protuberance (*see* figs. 2 and 4), and curving out from this on each side is a thick ridge of bone which corresponds to the superior curved line (fig. 4). Below the external occipital protuberance and

between the two superior curved lines the bone is much flattened (figs. 2 and 4), so that here it presents a concavity, and it seems as if one could map out the posterior part of the foramen magnum. The parietal bones do not articulate with one another (*see* fig. 3, P). They are dwarfed and their union is prevented by the fusion above them of the occipital (O) and frontal bones (F). They articulate above with part of the frontal bone and behind with the occipital bone. Below they are widely separated from the squamous temporal bones (T). The squamous temporal bone

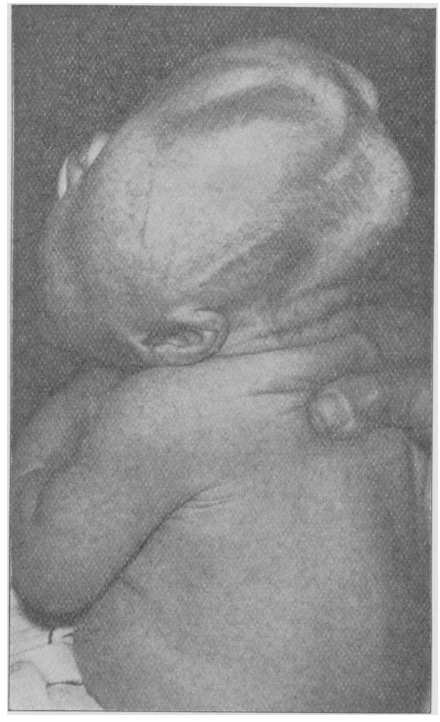

FIG. 2.

on each side (*see* fig. 3, T) is everted and looks downwards and outwards; it is small. There is therefore a distinct gap (*see* fig. 3, GAP) corresponding to what would normally be the anterior and posterior lateral fontanelles and the squamous suture. On each side above the zygoma (*see* figs. 1, 2, and 3) and in front of where the ears are attached is a soft round bulging mass consisting of a part of the brain protruding through a wide gap (GAP in fig. 3) between the frontal and parietal

bones above and the squamous temporal bone below. The mass does not pulsate and is probably hydrocephalic; an impulse is readily transmitted from side to side. The greatest circumference of the skull is 17 in. at the level of the supra-orbital ridges. From the root of the nose to the external occipital protuberance, directly backwards, the distance is $6\frac{1}{2}$ in. The greatest breadth is $6\frac{1}{2}$ in. The greatest length from the chin to the vertex is 7 in. From the root of the nose to the back of the skull is 5 in. The left optic disc is smoky-coloured, with a

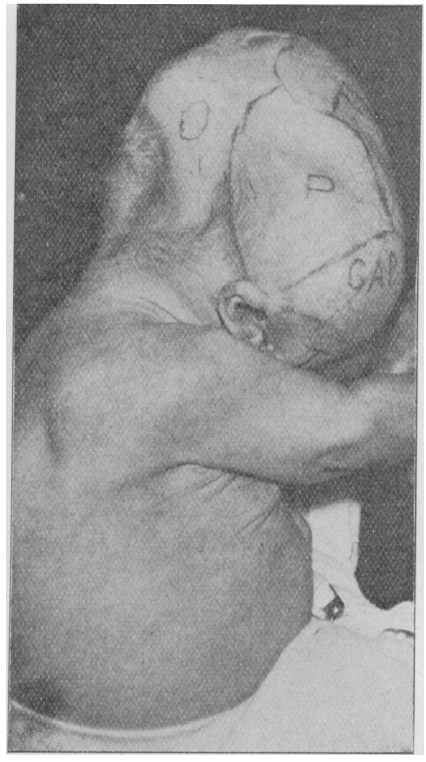

FIG. 3.

pale centre situated in an excentric position. The veins are large, the arteries small. The right optic disc is a light smoky colour with a central pale area. The veins are large, the arteries small. Both arms and legs are rigid and the knee-jerks are +.

There are also other congenital abnormalities as follows:—Hands: The middle and third fingers of each hand are webbed up to the base

of the nails. Feet: There are six toes on each foot, all directed inwards; the three inner on each foot are webbed. A skiagram shows a terminal phalanx for the supernumerary big toe (fig. 5), but no corresponding metatarsal bone as was shown in the sister.[1] There is a hernia the size of a marble in the linea alba midway between the umbilicus and the ensiform cartilage, the hole in the abdominal wall admitting the index finger, and another smaller rupture at the umbilicus. The heart is normal.

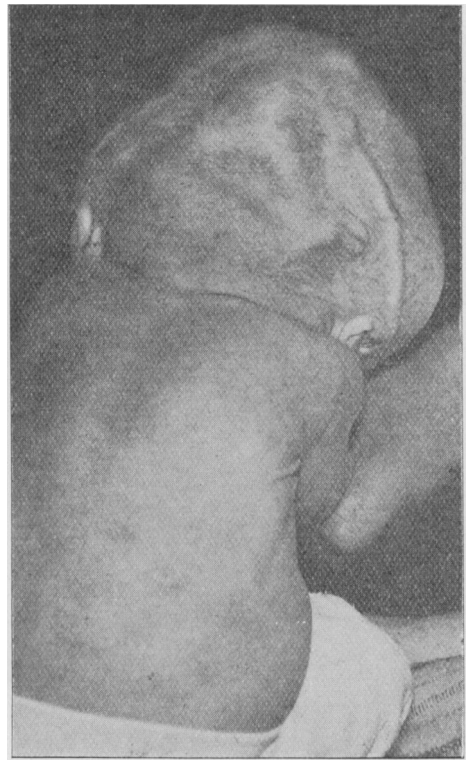

FIG. 4.

Eight years ago, at a meeting of the Society for the Study of Disease in Children,[2] held at the Belgrave Hospital for Children, then situate in

[1] *Reports Soc. for the Study of Dis. in Children*, Lond., 1901, i, p. 116, fig. 13.

[2] "Two Sisters showing Malformations of the Skull and other Congenital Abnormalities," *Reports Soc. for the Study of Dis. in Children*, Lond., 1901, i, p. 110.

Gloucester Street, I showed two sisters of this child, both of whom are now dead. At that time one girl (Dorothy) was aged 1 year and 11 months. Her skull was malformed but not to the extent seen in this patient. Her frontal bone rose wall-like, and it was on a level with the tip of her nose. Looked at from the front the forehead was pear-shaped, with the stalk of the pear at the root of the nose, and its base passed into a beak of bone. The sagittal suture passed forwards for some distance in the usual direction until within a finger's breadth of the coronal suture. Here a distinct, beak-like, bony prominence (mentioned previously) was encountered, which ran forwards for about 3 in. The beak was 1 in. broad at its widest part and $\frac{1}{2}$ in. at its narrowest. The sagittal suture bifurcated at the commencement of this bony

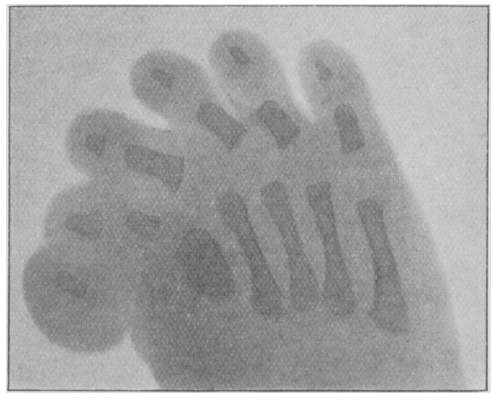

FIG. 5.

prominence, and on either side of it sutures could be felt. Behind the level of the coronal suture there was $\frac{3}{4}$ in. of beak-like prominence of bone, and in front the beak merged into the base of the pear-shaped frontal protuberance of the skull. In the child exhibited there is a central ridge 1 in. broad running from below upwards. The parietal bones on each side appeared to be made up of two bones, and the central beak was thought to be a Wormian bone. They were obviously quite different to the parietal bones in the case shown. The supra-orbital notches were large and sutures could also be felt running from them, differing in respect to the present case by the super-added sutures. The

[1] Op. cit., figs. 10 and 11.

posterior and postero-lateral fontanelles were patent. The skull was bulged on either side above the level of the zygoma, which ended about two fingers' breadth on either side of the median beak of bone. The root of the nose was flat, the upper lids were thrust down, and also the eyes. The veins in the fundus oculi were corkscrew-like and full of dark blood, the arteries were tortuous and perhaps a trifle small. The nose, lips, and tongue were blue and the extremities cold and cyanotic. She had a loud systolic bruit over the second left interspace, heard better over the left side of the chest than the right, and also audible in the great vessels of the neck. There was no thrill and no extension of the cardiac dulness to the right. The bruit was not heard in the back. *Hands:* The little and first fingers and the thumbs were partially webbed, and the ring and middle fingers completely so. Feet: There were six toes on each foot and all webbed except the little toe and that next to it, where the webbing was only partial. Only one bone could be seen in each of the little toes; in the three next, two bones; and in the two following toes, one bone each. *Ruptures:* There was a hernia about the level of the umbilicus admitting the tips of three fingers, and ¾ in. above that another rupture admitting the index finger. She died when aged 2 years and 9 months.

The other child (Florence) was aged 3 years and 10 months when I saw her. Her forehead was somewhat like that of her sister, but it did not overlap the tip of the nose. The skull was keel-shaped.[1] The keel commenced at the top of the forehead and terminated on a level with the parietal eminences. The back of the head was natural. The circumference of the skull measured 19 in. The eyes were prominent and small, with heavy upper lids; the lower lids were puffy. The nose was short and the bridge flattened. The upper lip was long and both lips were thick and pouting. The chin was short and receding. She looked old for her age. When she was cold her nose became blue and the face dusky-looking, but otherwise the circulation was not abnormal. There was a systolic murmur of limited conduction heard over the left base; the heart was not enlarged. Both optic discs were on the pale side and the veins were very tortuous. The hands showed the same deformity as in Dorothy. There were six toes on each foot, as with Dorothy; the big toes and the next were partially webbed, there was incomplete webbing between the little toes and the next, and the webbing was complete in the others; all the toes possessed two joints

[1] Op. cit., fig. 12.

excepting the big toe, which had but one; the metatarsal bones of four of the toes, starting from the little toes, were natural, the fifth appeared to be thickened, and the sixth were wedge-shaped, with their apices pointed forwards.[1] There was an umbilical hernia the size of a Tangerine orange. The cornea was imperfectly developed; at one portion its outlines were not elliptical, and the neighbouring sclerotic seemed to run into the cornea. This child went to an ordinary school, but she was mischievous. She set fire to herself when aged 10 and died from the accident.

The parents, both exceptionally strong and healthy-looking, have been married twelve years. There is no history of syphilis. Their third child (a boy, aged 7) is quite normal. Their fourth child was born two years later; it was premature, at seven months, and was born dead; the mother was told that it resembled the two eldest children in appearance. The fifth child is a boy, aged 3, who is also quite normal.

The deformity of the fingers can be traced to the mother's family, as her brother's child had deformed hands, but there is no history of cranial deformities to be obtained, and there is no obvious parental explanation for their occurrence. The mother states the child's head has got much bigger since its birth.

Mr. Henry Power, under the heading "Microcephalus and Proptosis,"[2] published a remarkable case of pyrgocephaly (tower skull) which was brought to St. Bartholomew's Hospital thirty-six hours after birth and died a month later. A drawing of the case, a cast of the head, and a section of the skull are in the museum of that hospital. In this infant proptosis was extreme and the skull markedly tower-shaped—that of a "blunt cone." The head was very short in its antero-posterior direction but of considerable height, especially in the frontal region. The orbit was extremely small—too small to receive the globe of the eye. The shape of the head caused the frontal convolutions of the brain and of the corpus callosum to be perpendicular in direction. The anterior fontanelle was present. Mr. Sydney Stephenson,[3] in a section on "Oxycephaly," draws attention to this condition of "tower skull" and gives an illustration of a boy under Professor Uhthoff with a proptosed eyeball which had been dislocated. He also gives two illustrations of a lad, aged 17, who came under his notice with a "tower skull" and very pronounced proptosis. But of all the illustrations of the condition that

[1] Op. cit., fig. 13.

[2] *Trans. Ophthal. Soc.*, 1874, xiv, p. 212 (illustrated).

[3] *Brit. Journ. of Children's Dis.*, ii, pp. 492-6.

I have seen, and of all the accounts that I have read, none can compare in grotesqueness of appearance and cranial deformity with the case that I exhibit. In my opinion any explanation of the deformity which relies upon premature ossification of sutures or of premature synostosis of the cartilage between the basi-sphenoid or basi-occipital bones is wide of the mark. To me it seems much more reasonable to assume a vice of development inherent to the cranial bones or their membranous precursors. I readily allow that vice of development is well enough as a phrase and that it is no real explanation, and that it is necessary to probe deeper than that for the intimate cause. But it is a better explanation than the others, which will not "hold water." There is no known mechanical explanation which could account for the gross exaggeration of the normal anatomical peculiarities of the occipital bone observed in this patient. Nor will such an explanation fit in with the deformity of the parietal bones of the patient shown or with the irregularities of the parietal bones found in his sister Dorothy. Although pressure *in utero* might theoretically explain the abnormal union of the frontal and occipital bones and the exclusion of the parietal bones from the scheme of formation of the infant's cranial vault, yet the giving birth to, by the mother, of children with normal skulls lends no support to the idea of there being defects in the maternal pelvis, which is the only structure which could reasonably be supposed to bring about such a condition. On the other hand, the coexistence of numerous and various associated congenital malformations, coupled with the maternal history, clearly indicates that the fault resides within the parental germ-cells, and that those of the mother are more likely to be at fault.

The child died on December 12, and a full post-mortem report will be presented to the Section at a future date.

A Girl aged 7 years showing Misplaced and Rudimentary Patellæ, Congenital Dislocation of the Hip, and Coxa valga.

By GEORGE CARPENTER, M.D.

MAUD F. W., an intelligent child, was born with deformities, her feet being "pointed" at the birth; the presentation was breech. The mother was a long time in labour, but no instruments were used. There have been two miscarriages, one just before the mother was pregnant