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ON TWO SPECIMENS OF ELEPHANTIASIS CONGENITA CYSTICA.

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ANTENATAL pathology presents no monster which, while preserving some semblance of the human form, deviates therefrom in stranger or more striking manner than the condition now under consideration. In figure 4, for instance, we see below two distorted legs, but no arms. Above the groins the head and trunk form a loose, shapeless sac, there is almost no indication of neck and shoulders. An irregular fissure stands for orbits and nasal fossæ, there are no eyes and the mouth is an irregular aperture. The whole is a travesty of the human form and features,

Monstrum horrendum, informe . . . , cui lumen ademptum.

In the other specimen the head has been destroyed by craniotomy and there is some attempt at upper extremities, but otherwise it closely resembles the companion specimen.

We take both these to be examples of the condition which has been called *Elephantiasis Congenita Cystica*. The best account of this will be found in Dr. Ballantyne's work on *Antenatal Pathology*, chapter xvii, see also his previous work on *The Diseases of the Fœtus*, chapter xiv., *et seq.*

The following description will be more easily followed if we point out that the malformation in question depends on three conditions.

1. Certain defects in the skeleton. These, however are not the

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essential thing. It is not as in spina bifida where the osseous defect causes the meningo-myelocele. The distorted contours depend upon disease of the soft parts.

2. There is general anasarca of the body.

3. The characteristic lesion consists in the formation of extensive, irregular cystic cavities in the subcutaneous tissue.

CASE I. This foetus was presented to the medical school by Mr. Sykes, of Barnsley, who has kindly furnished some notes of the case.

The mother, Mrs. K., aged 25, married 5 years, had two healthy children, then she had a miscarriage with a normal foetus. She then became pregnant for the 4th time and went to term. The abdomen was very large, and Mr. Sykes diagnosed a twin pregnancy by palpation. When labour came on the first child was expelled alive by natural efforts without difficulty. It was small but perfectly formed. The second child was not delivered until several hours later; the delay was due to the fact that there was hydrocephalus. Finally the head was perforated and not less than three pints of fluid escaped. Delivery was then easily completed and the mother made a good recovery.

Description of foetus. The head has been destroyed by craniotomy. The face cannot be found. The base of the skull and portions of the cranial bones and hairy scalp remain. The foetus is now about 10 inches long. A skiagram (fig. 1) was taken before dissection.

The trunk presents an ill-shaped fleshy mass which yields fluctuation beneath the integument in various parts, especially on the back. An umbilical cord of normal appearance hangs from the navel. There is no differentiation of shoulders. The left hand has 3 digits. Together with the lower part of the forearm it projects from the integument. By skiagram and by dissection it is easy to demonstrate that the scapula, clavicle, humerus and ulna of this side are present. The radius is wanting. The arm, as far as the elbow joint, is included in the general sac-like integument. On the right side no upper extremity appears externally. The clavicle, scapula, and part of the humerus are present however. The lower epiphysis of the humerus and all the forearm are wanting. The legs are present and are distorted by the swelling of the soft parts—the tibia and fibula are clearly seen in skiagram. There are three toes on each foot. The genitals are female. The anus is normal.

The foetus being laid prone a median incision was made down the middle of the back. On raising the integument extensive cystic cavities were seen in the subcutaneous tissue, and from these serous fluid escaped. The whole of the skin of the back was undermined by these cavities. For purposes of photography the skin flaps were held

FIG. 1. CASE I. Skiagram.

FIG. 2. CASE I.
Partial dissection of dorsal surface. Subcutaneous cysts well seen.
Scale of inches on right.

A little black horse-hair has been placed in Pharynx and Thorax, and under Costal Margin in Abdomen.

FIG. 3. CASE I.
Partial Dissection, Anterior View.

FIG. 4. Cass II.

apart and the cavities were packed with black horsehair. The photograph in fig. 2 was then taken. There was no spina bifida.

The monster was now placed on its back and a median incision was made from the upper end to the pubes (fig. 3). The two hypogastric arteries and the urachus were seen converging to the navel. The umbilical vein was not found. The abdomen was completely closed in by the diaphragm. The liver was absent. The intestinal canal commenced at the œsophageal foramen in the diaphragm, and was much convoluted, filling the abdomen. The stomach was not differentiated and there was little distinction between large and small bowel. The two oviducts and ovaries were well seen with remains of the Wolffian bodies, but the kidneys were not developed and no spleen was found. The thorax was open anteriorly, a V-shaped cleft extending from the neck to the level of the seventh rib. Apparently a portion of the sternum is present on each side, forming the limbs of the V, and to this the rib-cartilages are attached. The sternal ends of the clavicles are thus widely separated above.

The thorax was empty, the heart and lungs being absent. The tongue is well formed, and above this the glottis and pharynx are seen, but the trachea and œsophagus are represented by mere impervious fibrous bands. The specimen was decolorised and macerated from long immersion in spirit, and it was not possible to make a minute dissection or to demonstrate the relation of the great vessels.

CASE II. This specimen has been in the museum of the Leeds Medical School for an unknown period. No history has been preserved, and we are not aware that any description of it has been previously published; a good photograph is shown in fig. 4. The head is uninjured. The arms are wanting. The foetus is not available for dissection. Long immersion in spirit and suspension by the head have doubtless altered its appearance. It is a remarkable object, and except as regards the lower extremities and the umbilical cord, presents little resemblance to a human foetus. The head and trunk form a loose flaccid sac. A skiagram shows the vertebral column, pelvic bones, thigh and leg bones well developed; one femur is fractured. The feet seem well developed, and there are five toes on each foot. Traces of the ribs can be seen and certainly one clavicle, but the upper extremities are wanting. Unfortunately the skiagram yielded by this foetus is not sufficiently defined for reproduction here. It closely resembled fig. 1, except as regards the arm. The genitals are female; anus absent.

The face is extremely defective. It shows an irregular aperture for the mouth. The two orbits run into the nasal fossæ on account

of the absence of the nasal process of the superior maxilla. The orbits are empty. The septum nasi and inferior turbinate bones are present. The ears are not found.

This specimen is, we think, clearly of the same nature as Case No. i. Whether it is also acardiac cannot be stated in the absence of dissection; probably it is acardiac.

I am indebted to Mr. R. W. Taylor, one of our students, for the photographs of Case i. and to Mr. Oliver, of the Infirmary, for that of Case ii. The skiagram is by Messrs. Reynolds and Branson.

Summary. The first foetus may be described as an acardiac monster—a parasitic twin. It was affected with hydrocephalus and cystic elephantiasis and general dropsy. It had developmental defects in anterior thoracic wall and in extremities, with great defects in internal organs; heart, lungs, liver and spleen being absent; kidneys rudimentary, and alimentary canal imperfectly developed.

The second had also cystic elephantiasis, absent upper extremities and rudimentary development of face and skull. History wanting. Internal dissection not made.