A SPECIMEN OF THE SO-CALLED SIREN-MALFORMATION (SYMPUS, SYMELIA).

By JOHN H. TEACHER, M.B., C.M., and JOSEPH COATS, M.D.

From the Pathological Laboratory of the University of Glasgow.

THE malformation, of which the subject of the present paper is an example, receives its most usual name of *Siren-malformation*, or even *Sirenia*, from the resemblance in general form to the mythological inhabitants of the sea, whose fatal charms were supposed to be exercised on sailors. The specimen is a typical one, and occupies a middle position between the more extreme and less pronounced forms.

For the specimen we are indebted to the kindness of Dr. John Adams of Glasgow, who gives the following account of the circumstances connected with the birth of the child :—

“The mother is a healthy woman about 32 years of age, with a good family history and also a good record of previous health. She has borne three children at the full time, all well developed and healthy at the time of birth. She has had no miscarriages. In the course of the present pregnancy, no extraordinary feature of any kind occurred. Movement was felt when the pregnancy was advanced about $4\frac{1}{2}$ months. There is no story of any maternal impression, nor of any fright, nor of any misshapen individuals being seen or imagined during the whole period of gestation.

“Labour began on a Friday afternoon with slight pains at long intervals—an hour or so at first. These pains continued till the Monday morning, when I was called about three o’clock to find the woman running about the house and not seemingly in any great distress. She was getting anxious, as she was not aware of making any progress. The membranes were said to have ruptured during Sunday afternoon.

“She was put to bed and examined. The presenting part gave the idea at first of a breech, but further investigation did not confirm this, as no furrow between the buttocks or anal orifice could be discovered. The ease with which a foot could be reached led to the conclusion that the presentation was one of a knee. The pelvis being roomy and the patient’s strength good, it was resolved to attempt delivery without turning. With the forefinger of the right hand in the flexure of the knee, and with the left hand exercising friction and pressure on the abdomen, strong pains were induced, and the product of conception was expelled in about fifteen minutes. The fœtus had evidently been dead for three or four days.

“The woman made a good recovery.”

The fœtus as received at the laboratory was described and figured

before any dissection was attempted. The viscera were then examined and removed, and afterwards part of the skeleton was dissected out. The external parts were now restored as nearly as possible, and the foetus as a whole and the parts of the skeleton are preserved in the museum of the Western Infirmary, Glasgow.

The general aspects of the foetus are shown in Fig. 1, which

FIG. 1.—View of foetus from behind. The limbs coalesce and end in a single foot, having seven distinct toes and two coalesced ones higher up. The two great toes are at the outer aspects of the foot. The dorsal aspect of the foot is that shown, and it faces backwards. There is one rounded buttock with a large anal opening. The left hand has four fingers.

represents a posterior view. The trunk ends in a single rounded buttock, near the upper part of which is the anal aperture. The two lower limbs are coalesced into one broad member, which tapers downwards and ends in a broad foot, not unlike a fish's tail, furnished with nine toes. The left hand has only four fingers.

Making the examination more in detail, it was found on handling the foetus that it could be very readily doubled up anteriorly, there being

exceedingly free movement between the trunk and lower extremity. This movement was obviously not at the hip-joint, but higher up, as the pelvis moved with the lower limb. The seat of this loose connection was found on dissection to be the sacro-iliac articulation.

The aperture in the upper part of the buttock was obviously the anus pushed backwards and upwards. It admitted a good-sized quill, and a probe could be passed several inches into the rectum. Immediately above the anus there is a small dimple in the skin (see Fig. 1), and the tissue here was cicatricial in appearance. The dimple seemed to be in connection with the tip of the coccyx.

Viewed anteriorly no proper genital organs were visible. The only indication was a small projection about the size of a good-sized pin's head, situated in the middle line, 6.5 cm. ($2\frac{1}{2}$ inches) below the umbilicus.

The single lower limb has evidently arisen by a coalescence, in some places by no means complete, of the two ordinary limbs. But the coalescence has not occurred by the aspects which are nearest one another in the fully developed legs. It is as if the limbs before coalescence had been so placed that their outer aspects were in the middle line behind and had united and become fixed there. Thus the normal outer borders of the limbs are merged together in the middle line, and their structures partly suppressed, while the inner have become the outer borders. In this way the hip-joint is behind; the knee is reversed and flexes forwards instead of backwards. The plantar aspect of the foot presents forwards, and the great toes are at the outer borders of the foot.

Entering more into detail, it could be felt before dissection that the double femur was, especially at its lower part, unduly broad, and that it had a double lower extremity. The single foreleg was attached to the right half of the coalesced femur, and there was an abortive left foreleg, 2 cm. in length and tapering to a point, attached to the articular surface of what represented the left femur.

The foot was loosely articulated at the ankle, and its plantar aspect presented forwards. The foot was an almost direct prolongation downwards of the leg. It ended in a row of seven toes (see Fig. 1), of which the two outer are the great toes, and, of the remaining ones, three seemed to belong to the right foot and two to the left. In addition there were visible, on the dorsal aspect, two small coalesced toes, forming a broad but short double toe with two small nails. There are thus, in all, nine toes represented. The width at the ankle was 2 cm., whilst across the outspread toes the breadth was about 4 cm.

The upper limbs were well formed, with the exception that the thumb of the left hand was wanting, and the only indication of it was a small projection, oval in shape and the size of a pin's head.

On opening the body the following conditions of the intestine were found, there being three complete occlusions of the calibre of the gut, two of them with entire interruption of the continuity.

Taking the structures from below upwards, it was found that the anus communicated with a well-developed rectum, and that the descending colon was apparently normal. This part of the intestine terminated above in a rounded blind end, which was situated about 16 cm. above the anus. Examining further, the intestine was resumed on the right side by a bulky blind end, which represents the upper part of the ascending colon or transverse colon. To the apex of this a narrow tendinous band was attached, representing collapsed and occluded ascending colon. This being traced downwards there comes first a small finger-like diverticulum, 1.5 cm. in length, and then a narrow caput cæcum with vermiform appendage and ileo-cæcal valve. These are about 23 cm. from the upper blind end of this part. From the ileo-cæcal valve the small intestine was only traceable upwards for a distance of 11.5 cm., and here it suffered a complete interruption with a rounded blind end. There were still abundant coils of small intestine, but their relations could only be made out by tracing the parts downwards from the stomach. The stomach itself was found to be normal, and the small intestine was continuous through the various coils till it ended in a dilated and somewhat infiltrated piece of intestine.

It will thus be seen that the great intestine has been completely cut across, apparently in the situation of the transverse colon, possibly with removal of a portion of the colon. The two ends produced by the lesion have formed rounded bulbous extremities. Then, also, the small intestine a short distance above the valve has undergone a precisely similar interruption. In addition, the ascending colon has had its calibre abolished over a considerable extent, but without absolute severance of its continuity.

The two kidneys were found in their normal position. The testicles were also found lying in the abdomen and apparently normal. The liver and spleen were also apparently normal.

The *skeleton* was removed from the second lumbar vertebra down to the ankle-joint, and, without further dissection, the following points were noted (see Figs. 2, 3, and 4). The sacrum was very loosely articulated to the iliac bones, and it was due to this fact that the pelvis and lower extremity could be so freely doubled up on the abdomen. The most prominent peculiarity of the pelvis is the fact that the ischial bones have coalesced across its floor (see Figs. 2 and 3). Behind the coalesced ischial bones the rectum passed out, being thus pushed backwards and upwards. This accounts for the peculiar position of the anus. The coccyx and sacrum were tilted backwards (Fig. 2).

A more detailed examination of the portions of the skeleton removed gives the following results, which are illustrated in the sketches on the opposite page.

The parts removed are these: the last three lumbar vertebræ, the

pelvis, one large bone representing both femora, the tibia and fibula of the right leg and both patellæ.

The three vertebræ were found to be normal in front. They are closed behind, so that there is a complete spinal canal, but the laminae are cartilaginous, and there are no spinous processes. Fig. 3, which

FIG. 2.

FIG. 3.

FIG. 4.

FIG. 2.—Part of the skeleton seen from behind. The broad femur has a double head, which articulates with the pelvis behind. The patellæ are shown, the left looking backwards, the right backwards and outwards.

FIG. 3.—Three lumbar vertebræ and sacrum seen from behind. The arches of the vertebræ are incomplete, and there are no spinous processes. In the sacrum the vertebral canal is completely open.

FIG. 4.—Pelvis seen from the front. The ischial bones are coalesced across the floor of the pelvis. The pubic bones are also coalesced and are projected forwards.

represents the posterior aspect of the vertebræ and sacrum shows this. The sacrum, as shown in Fig. 2, is bent backwards at right angles to the vertebral column, and is also slightly twisted towards the right side. In the sacrum (see Fig. 3), the vertebral canal is completely open, two or three tubercles at each side representing the laminae.

The iliac bones seem to be well developed. They diverge widely,

and they are somewhat rotated, so that their anterior borders look directly downwards. The lines of junction with the ischia are well marked in front.

The ischial bones are markedly altered, being transformed into a thick bar of bone which crosses the pelvis nearly parallel to the anterior surface of the sacrum (see Fig. 4). From this project forwards three processes, which meet in the middle line in front. The two lateral ones, which represent the horizontal rami of the pubic bones, lie higher than the third, and form a rounded projection anteriorly. The third, which seems to represent the two descending rami of the pubic bone united into one, is in the middle line, and passes from the middle of the coalesced ischia to the pubis in front. This median process was separated from the lateral ones on either side by a small aperture, and these apertures represent the obturator foramina. To the lower border of the median process was attached a rudimentary penis.

Between the bar of bone representing the ischia, and the sacrum, is the pelvic outlet, which is much narrowed from before backwards. It measured 2.25 cm. transversely, and from before backwards .5 cm. at the left, and .7 cm. at the right side (see Fig. 2).

On the posterior aspect of the pelvis, just at the junction of the ilia with the transverse bar, are two acetabula, looking backwards and slightly downwards. The synovial cavity of each is quite distinct.

The two pubic bones are coalesced in front and projected considerably forwards (see Fig. 4). The horizontal rami are separately distinguishable, but the descending have coalesced into the single process, which, as already mentioned, joins the bar formed by the coalesced ischia. The infrapubic arch is thus abolished.

The two femurs are represented by a single shaft, which is bifid above and below (see Fig. 2). Above, there are two distinct heads articulating separately, two necks, and two great trochanters. The shaft itself is markedly flattened and broad, measuring 1.2 cm. at its narrowest part.

The lower end bifurcates a fifth from its extremity, and there are two distinct and separate articular ends, each having the proper surfaces for the knee-joint and its own patella. The position of the articular surfaces in relation to the normal, is altered in accordance with the rotation of the limb already mentioned. This is indicated by the position of the patellæ (see Fig. 2), the left patellæ looking almost directly backwards, and the right backwards and outwards. The right knee-joint is completed by the tibia and fibula. The left was formed by the aid of a bone which was left in the body. There is a fibrous separation between the two ends of the femur.

The tibia and fibula are normal except at the lower extremities, which are very slightly altered so as to articulate with the deformed foot.

Regarding the bones as a whole, the extreme flexibility of the lower extremity forwards is explained by the looseness of the sacro-iliac

synchondroses, by the position of the hip-joints, which prohibits backward movement but allows of considerable flexion forward, and by the reversal of the knee so that it flexes forwards instead of backwards.

REMARKS.

The specimen described above conforms in most respects to the cases of a similar lesion described by various authors. There are differences in the degree of coalescence of parts in the recorded cases, but in their general features and in the kind of defect in the pelvis and leg there is a close similarity.

Taking the classification of such malformations adopted in the exhaustive work of August Förster,¹ which, with differences of nomenclature, is virtually that of Geoffroy Saint Hilaire,² we find that our specimen belongs to the general class of *Sympus* or *Sympodia*, signifying a coalescence of the lower extremities. Geoffroy Saint-Hilaire uses the more general term *Symèle* (μέλος, a limb). The class is divisible into three groups which, to some extent, merge into each other but are sufficiently distinct for practical purposes. There are first the cases in which the defect is greatest; the foot is absent altogether, and the leg ends in a rounded or tapering extremity. This is the *Sympus apus*. Geoffroy Saint Hilaire limits the term *Siren* to this form, and designates it *Sirénomèle*. The second group is that to which our case belongs. There is such a coalescence that there is only one foot, although the toes may be more numerous than those of the normal foot. This is the *Sympus monopus*, or the *Uromèle* (οὐρά, a tail). In the last group there are two distinct feet, and this form is called *Sympus dipus*, or by the general term for the whole group, *Symèle*.

The peculiarities which are common to the whole group, and which give it its unity, may here be enumerated, as they are illustrated in our case. It seems to us that the central item in the defect is the condition of the pelvis and especially of the ischia. In Förster's very careful descriptions of the three forms, the coalescence of the ischial bones is the only absolutely constant lesion of the skeleton. In our case the two ischial bones form a transverse bar across the pelvis, and this seems to be the regular condition. Even in the least advanced cases this feature is present. There is a case of *sympus dipus*, for example, related and illustrated by Cruveilhier,³ and given also in the Atlas to Förster's work, in which all the bones of the leg and foot are doubled, and the only coalescence is between the two calcanei; but the pelvis, as appears from Cruveilhier's description, and his most careful illustration, is very much like that in our case. The two ischia are united, and the

¹ Förster, "Die Misbildungen des Menschen, nebst Atlas." Jena, 1865.

² Geoffroy Saint-Hilaire, "Histoire générale et particulière des Anomalies, avec Atlas," Paris, 1832.

³ Cruveilhier, "Anat. Pathol.," livr. 40, plate vi.

pubic bones are pushed forward prominently, in a similar position to that shown in Fig. 4.

Another constant feature is the altered rotation of the limbs, which is so fully illustrated in our case, and this connects itself directly with the central lesion just mentioned. It seems to be a common feature that, when the bones of the leg coalesce, they are united by their external borders, which have been rotated backwards and inwards. Even in cases of dipus, where the whole of the bones of the leg are doubled, the fibulæ are to the inside and the tibiæ external; and the great toes are at the outer margins of the double foot. This peculiarity is to be related to the coalescence of the ischia, which we have regarded as the central lesion. The coalescence of the ischia, in the middle line behind, displaces the two hip-joints backwards, so that they approximate to each other, or even coalesce posteriorly. In this approximation of the acetabula, there is necessarily a rotation of these articular surfaces. The acetabula, instead of looking outwards and forwards on each side of the pelvis in the normal fashion, will, in proportion to their approximation to the middle line behind, come to look more and more backwards. Following on this, the femora will be similarly rotated, so that their articular surfaces, instead of presenting, as in the normal hip-joint, inwards and backwards, will come to project according to the degree of the lesion, more and more directly forwards. This will bring into close proximity the posterior and outer aspects of the femurs, and cause them to be fixed in this rotated position.

The degree of the lesion will be determined by the extent of the coalescence of the ischia, and the consequent approximation of the acetabula behind. There may be such an approximation of the acetabula as to make only one joint cavity, and that a shallow one; and this coincides with the extreme forms of the defect in symphysus. On the other hand, there are the various degrees of two acetabula with united femora (as in our case), and a single foot, on to those in which the acetabula, being considerably separated, the bones of the leg and foot are completely doubled. It will be obvious that it is this rotation of the femurs, and their approximation by their posterior and outer borders in the middle line behind, that brings about the rotation of the leg as a whole; the abnormal flexion of the knee-joint, and the position of the great toes externally, and the little toes internally.

Lesions of the terminal parts of the intestinal and urino-genital apparatus are apparently constant in all forms of symphysus. It might be expected that the coalescence of the ischia, directly across the position of the anus, would be liable to abolish that orifice and the last part of the rectum. It has not done so in our case, but this seems to be an entirely exceptional circumstance. Imperforate anus is stated as a constant feature by Förster, who says, in his account of the whole group, "Der After fehlt stets." In our case the anus was pushed backwards, but the rectum and outlet were well developed. The three in-

terrutions of the calibre of the intestine, however, were equally effective in rendering the continuance of life impossible, even if the child had been born alive.

The urino-genital organs are also in all cases interfered with. There may be no appearance of external organs, or a mere trace as in our case, or an approximation to the normal genitalia, but it is stated that there is never any opening of the urino-genital passages externally. Whether this is liable to exception, as in the case of the anus, remains to be seen. It is stated by G. Saint-Hilaire that the kidneys, at least on one side of the body, are always involved. In this respect also our case is an exception, as both kidneys and both testicles were present and apparently unaffected.

We come now to the question of *the origin and causation* of this form of monstrosity. In view of what has been said as to the central point of the defect being in the floor of the pelvis, we have to consider what kind of lesion may have produced it, and at what period of development. The lesion is not a mere error in development. It is a lesion, having certain definite and constant characters, which are all traceable to a defect in a particular part of the foetus.

The lesion must have originated at an early period of utero-gestation. The formation of the buds which go to form the lower limbs must have just begun. There was no differentiation as yet of the individual bones in any part of the limb, except, it may be, of the digits. The buds forming the lower limbs are placed at first as flaps against the lower part of the body of the embryo. These flaps, at first, represent little more than the foot, whose elements are the first to appear, and they are applied to the body, so that the great toe is towards the cephalic end of the foetus, and the little toe towards the caudal end. In this early period, should some agent acting on the posterior parts of the embryo, where the primordial pelvis is situated, cause an approximation of the limb buds behind, then the two buds would approximate by what in adult life are their outer borders; that is to say, by the parts which the little toes denote. This may, perhaps, be more clearly understood if the two hands be placed on either side of the pelvis, with the palms against the body and the thumbs upwards. A defect in the pelvis would cause the approximation of the edges of the little fingers, greater or less in degree, according to the amount of the defect.

But a mere approximation of the parts is not in itself sufficient to cause coalescence, and it is not sufficient to cause those actual defects in the structures which we have seen to exist. It seems to us that the state of matters implies that some injury at this early period has not only removed or destroyed some of the parts, but has, as in the case of raw surfaces in the adult, induced a tendency to coalesce. It looks as if a bit of the floor of the pelvis had been, as it were, pinched out, so that the germinating tissue was broken up, the epiblastic or cuticular layer being removed, so that it acquired a tendency to adhere

to the structures opposed to it. The injury to the tissue in some cases involves the limb buds themselves, as distinguished from the pelvis, so that there may be great destruction of them, and a mere aborted extremity formed, as in *sympus apus*. Or the limb buds may be left nearly unaffected, and their whole parts, at least so far as the skeleton is concerned, may be perfectly developed, as in some cases of *sympus dipus*. All this implies that the lesion occurs when there is little more than the limb girdle and the first buds present. The formation of the limb girdle, which afterwards forms the pelvis, occurs about the same time as that of the buds, namely, about the fourth week of uterogestation.

That the lesion is something in the nature of an injury is evidenced further by the condition of the intestine, and by the occasional further defects in the urino-genital organs, not merely the interruption of passages, but such circumstances as the absence of the external genitalia, and various defects of the kidneys.

It may be further remarked that the injury is essentially one of the middle line, producing a malformation which is virtually symmetrical, although not in every case absolutely so. It is curious that in our case, for instance, there is a doubling of the lower end of the femur and upper end of the tibia; but, on the one side, the greater part of the foreleg has, as it were, been picked out.

The question next occurs as to the nature and mode of production of such an injury at an early period of foetal life. Cruveilhier¹ suggested that the action of lateral pressure on the posterior parts of the embryo might produce it. But it seems to us that lateral pressure could scarcely produce such a definite lesion, limited in its primary seat to the middle line; nor would it explain what to us is so obvious as the principal fact, the removal or suppression of certain structures. Dareste² agrees with Cruveilhier that the cause is to be found in pressure, but does not see how external pressure could cause the singular rotation of the limbs, which is so characteristic. He claims that he has found the true explanation, in an arrest in the development of the posterior part of the amnion, which he calls the *Capuchon caudal*. By this arrest the capuchon is not expanded below the embryo, but rests on the latter instead of separating from it. In consequence of the constriction by this part of the amnion, the limb buds are turned backwards, and come to be placed towards the dorsum of the embryo, approximating by their external aspects, which have now become internal. This looks more like a correct explanation, but it does not by any means account for the defect in the pelvis, which seems to us to be a primary fault. Nor does it explain, as Dareste is careful to point out, the defects in the urino-genital and intestinal tracts, which are such constant concomitants. We regard it as probable that defect of

¹ Cruveilhier, "Traité d'Anat. pathol.," i. 311.

² Dareste, "La production artificielle des Monstruosités," 2nd ed., 1891, p. 420.

the amnion, either by way of simple non-expansion of the caudal end or abnormal adhesion, so as to interfere with the growing structures, is the cause of the malformation.

The peculiar position of the coalesced limb has been frequently referred to. In this connection it is to be remembered that, in the process of development and expansion of the foetus, both the upper and the lower limbs undergo a process of rotation. The lower limbs appear as two lateral buds, which are applied to the sides of the body. They are so applied that the plantar aspects, or what will become the plantar surfaces, face inwards or even slightly forwards, and are in contact with the body. The primitive toes are spread out, and are counted from above downward, the great toe being the first and uppermost. In the normal process of development, the limb-buds suffer a rotation, so that the plantar surfaces ultimately face downwards and the great toes inwards, although at the time of birth, and even for a time after it, the soles of the feet frequently look inwards, and the great toes upwards.

The lesion under consideration will altogether prevent this rotation. The parts which become the outer borders of the limbs, by the defect in the middle line, are brought into contact and adhere. The coalesced limb is thus fixed, with the outer borders adhering in the middle line, and the limb grows out with the plantar aspect facing forwards, the great toes outwards, and the little toes, more or less coalesced or suppressed, internal.