Spontaneous Intrauterine Decapitation

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Understanding of the significance of congenital malformations advances only slowly. One of the most difficult to explain is acephalia—complete absence of the head—of which an example is described here.

True acephaly where the whole head is missing from an otherwise well-developed foetus must be distinguished from anencephaly, of which the features are well known, and the acephalic acardiac variety of reduced twin (Willis, 1962). Here, though no head may be visible, some of the organs and a brain in much reduced form may be buried in the amorphous body.

Case Report

(Case I in Table)—The mother, aged 24, has given birth to three infants. The first, a girl, born 1961, weighed 1871 g. (4 lb. 2 oz.). The third, a boy, weighing 2919 g. (6 lb. 7 oz.) was born in 1964. Both were entirely normal.

Her second pregnancy in 1963 was also uneventful, until she was admitted in the second stage of labour at approximately 33 weeks' gestation. One hour later a slightly macerated headless male foetus was delivered by breech extraction. Because of a retained placenta the uterus was explored under general anaesthetic and the placenta was removed manually. At the same time a search was made for the missing head, but no trace was found. The puerperium was without complication.

The foetus (Fig. 1 and 2) weighed 750 g. Between the shoulders, the neck formed a conical stump surmounted by a leaf-shaped dark bluish-red shiny area. Maceration was moderately advanced and sheets of epidermis were separating from the limbs, abdomen, and perineum. Loose epidermis formed a 'collar' about 1 cm. wider than the smooth 'skin' over the stump.

The trunk and external genitalia were normal. There was bilateral talipes equinovarus and a similar deformity of the hands manifested by fixed flexion of the metacarpophalangeal joints, with hyperextension of the terminal interphalangeal joints.

At the neck from before backwards were the following features: (a) A tiny pore, partly covered by a thin fold of tissue and leading into the oesophagus (Fig. 3). (b) A slit flanked by two folds enclosing the laryngeal

cartilages and opening into the larynx and trachea. (c) A tiny tuft of hair over the vertebral canal.

The sterno-mastoid muscles were absent. Dissection confirmed that the larynx was present and led into the normal trachea and bronchi. The lungs were well developed and consisted of the usual lobes, two on the left side and three on the right.

The oesophagus was normal below the small opening on to the neck. The stomach and intestinal tract were completely normal. The liver was soft due to advanced autolysis. The thyroid gland and thymus were normal. No suprarenal glands were found. The testes were both within the abdominal cavity. The genito-urinary tract was normal. The heart, aorta, and pulmonary vessels were normal, as were the umbilical vessels. The placenta, 12 cm. in diameter, showed areas of scarring, but not of exceptional size. The umbilical cord showed no knots or other abnormality. The membranes were very ragged. Histological examination of the lungs showed no evidence of inhaled amniotic squames. Autolytic changes were advanced.

An x-ray picture of the whole skeleton (Fig. 4) showed normal development consistent with the length of gestation. Apart from the talipes already mentioned, the only abnormality was in the upper cervical region.

Separation of the head had occurred between the 1st and 2nd cervical vertebrae. In addition, the bodies of the 2nd and 3rd vertebrae formed a block and the arches were unfused.

Comment

Few examples of this rare malformation have been recorded. The first, quoted by Kohler (1962) was mentioned by J. B. van Helmont (1577-1644) in his '*Ortus Medicinae*' (1652). Amongst his examples of intrauterine amputation is an account of the birth of a decapitated infant with a bleeding neck, to a woman who was watching public executions of rebels against the Spanish overlords.

The rarity of this type of defect can be judged from the fact that of the 55 headless monsters described by Tiedemann (1813), amongst those of which full anatomical details are given, there is not a single acceptable case of apparent decapitation. All those presented are varieties of acardiac acephalic twins. In two cases where only an external description is available and which include fully

Received February 6, 1967.

FIG. 1.—Ventral view of foetus described (Case 1).

FIG. 2.—Dorsal view.

FIG. 3.—The stump of the neck, showing orifices leading into the larynx and oesophagus.

developed arms and legs, the possibility remains. That described by Marcus Mappus—quoted by Tiedemann, illustrates the difficulty.

Marcus Mappus in his '*Historia Medica de Acephalis*' (1687) gives a review of travellers' tales about headless tribes and mentions several medical cases of various types, ending with a brief description

of two foetuses. One is an amorphous monster. The other is of great interest in that it was a twin born at 6 months' gestation, the first of the pair being normal. If the artist is to be trusted, and the author assures us that the drawing is exact, it was a well-developed foetus (Fig. 5a and b) with a smoothly rounded neck stump without scars. There was a mid-line fissure over the sternum from which two nodules protruded. Probes could not be passed through the fissure—a fact particularly noted, since Mappus was using these cases to settle the argument as to whether the foetus *in utero* was nourished orally or via the umbilical cord. Only 3 toes were shown on each foot. The umbilical cord was knotted and the distal end was shrivelled. Unfortunately permission for even limited dissection was refused.

Another example, of which only a very brief description is given, was demonstrated at a meeting of the Berliner Medizinische Gesellschaft by Landau (1908). This was a 17 cm. long foetus, born at 5 months' gestation. There was bilateral club-foot, atresia of the genitalia and anus, with micromelia of the left arm. It was headless, but fixed to the placenta near the attachment of the umbilical cord was a slightly shrunken head with a short neck. The head was the size of a hazel nut, so that evidently bodily development had continued after decapitation. There is no account of the internal organs.

A headless female infant (Case II—Table), weighing 950 g. and born alive at 37 weeks' gestation, is described fully by Klöppner (1950). It was the

FIG. 4.—X-ray film of Case 1.

FIG. 5.—*The acephalic fetus described by Marcus Mappus.*

TABLE
Clinical and Anatomical Data of Present Case and 3 Others

	Case I	Case II: Klöppner (1950)	Case III: Ehrhardt (1956)	Case IV: Benešová (1960)
Level of decapitation	Above 2nd cervical vertebra; fusion of bodies of 2nd and 3rd cervical vertebrae	Above 4th cervical vertebra	Above 1st cervical vertebra	Above 2nd cervical vertebra which lacked transverse processes
Skeletal abnormalities	Bilateral club-feet; club-hands	Bilateral club-feet	Bilateral club-feet	Bilateral club-feet
Neck scar—Skin	Cervical stump; oval skin defect; smooth bluish area over stump	Cervical stump; double skin defect	Cervical stump; possible skin defect	Bluish smooth skin over cervical stump
Oesophagus	Patent	Partially blocked opening on to stump	Patent	Oesophagus became a solid cord above D1
Larynx	Patent	Partially blocked larynx (or trachea only) opening on to stump	Patent	Trachea became a solid cord above D1
Vertebral column	Marked by tuft of hair	Covered by soft brown tissue	—	Narrowed spinal cord; canal covered by fused skin and dura mater
Cardiovascular	Normal	Normal, including carotid arteries	Normal	Absent carotid arteries
Endocrine system— Thymus Thyroid	Present Normal	Small Not dissected	Large Not described	Normal Absent; parathyroids absent
Suprenals	Absent	Very small	Normal	Very small
Sex	Male	Female	Female	Female
Weight	750 g.	950 g.	2000 g.	2000 g.
Gestation	33 weeks	37 weeks	41½ weeks	—
State at birth	Macerated; time of death not known	Lived 20 minutes	Macerated; foetal heart heard up to 8 days before delivery	Lived 10 minutes
Maternal features	2nd out of 3 pregnancies; retained placenta	3rd out of 5 pregnancies; polyhydramnios; retained placenta	3rd pregnancy; membranes ruptured 4 mth. before delivery	5th pregnancy; intermittent bleeding up to 4th mth. of pregnancy

result of the third of the mother's five pregnancies. At birth the infant moved its limbs in response to stimulation. A visible and audible heart beat continued for 20 minutes before the foetus was declared dead. No respiratory movements were observed. The neck ended above the 4th cervical vertebra. The stump was covered by soft brown tissue, surrounded by bluish-red smooth skin. Two oval defects in the skin separated by a narrow bar were present. From the anterior area two pits on the surface led into the oesophagus and larynx or trachea (the demands of the museum precluded a satisfactory dissection of this site). The lumina were partially blocked. Again bilateral club feet were observed. The thymus was small, and the suprarenals very small and similar to those seen in the anencephalic foetus. The thyroid region was not dissected out.

The mother had complained of continuous abdominal pain in early pregnancy. Polyhydramnios had been present later and manual removal of the placenta was needed after the delivery.

Ehrhardt (1956) recorded a similar headless female infant (Case III—Table), born at 41½ weeks' gestation, weighing 2000 g. The foetal heart could be heard up to eight days before delivery, though the membranes had ruptured 4 months previously and there had been intermittent loss of liquor since. There had previously been a triplet pregnancy and an abortion followed by endometritis. She had received progesterone ('proluton') every 2½ weeks up to the fifth month of the present pregnancy. The foetus resembled the other two in that bilateral talipes equinovarus was present. The neck ended in a conical stump over which there was a possible defect in the skin. Maceration made this point uncertain. Separation had occurred between the base of the skull and the first cervical vertebrae. The oesophageal opening was patent and the laryngeal cartilages bordered the second opening. The thyroid gland was not described, but it was emphasized that the suprarenal glands were of normal size and the thymus was also large. The placenta was normal, but the membranes were ragged, though no amniotic bands could be detected.

Another headless female infant, weighing 2000 g. and alive at birth, was reported by Benešová (1960) (Case IV—Table). This was the fifth child of a 40-year-old woman. It survived for 10 minutes after birth. Bilateral club-feet were present and the neck stump was covered by smooth bluish skin. The head and first cervical vertebra were absent, but the remaining vertebrae were well developed. The surface resembled a healed operation stump. Like Klöppner's case, the suprarenals

were very small and the thymus was also smaller than normal. It differs from the rest in that the trachea and oesophagus both ended in solid cords of scar tissue above the level of the first thoracic vertebra, the cords eventually disappearing amongst neck muscles. The thyroid gland and parathyroids were completely absent, and another significant anomaly was the absence of any cervical branches of the aortic arch; two large branches only were present and no carotid arteries were found. The vertebral canal was roofed by dura, which at this point was fixed to the overlying skin. The spinal cord narrowed suddenly above the second cervical segment. During pregnancy there was intermittent bleeding in the early weeks, which was succeeded by the development of polyhydramnios in the fourth to fifth months.

For ease of comparison the main features of all 4 recent cases are presented in the Table.

Discussion

The fact that these headless foetuses not only continued to develop, but that two of them were actually born alive, while a third survived to within a week of parturition, is remarkable.

Acephalic and acardiac monsters in which the abnormality obviously dates from early in the embryonic period are well known (Boulgakow, 1926; Gladstone, 1905; Willis, 1962). They are invariably twins—the other one usually being normal. In these monsters bodily development and particularly that of the arms is usually severely disturbed; the majority lack a head and arms except in a rudimentary form. Even those most fully developed usually show defective digits and limbs (Tiedemann, 1813). Admittedly the absence or gross maldevelopment of the heart is an important factor. Pharyngeal and foregut derivatives are usually entirely missing. These features are in striking contrast to the cases being discussed here.

In each case the completeness of the thoracic viscera and neck structures up to the point of separation makes it evident that these are examples of decapitation of a well-formed head and not of primary maldevelopment. The perfect structure of the neck organs and other foregut derivatives makes it inconceivable that separation of the head took place in the embryonic period—the first eight weeks of gestation during which the rudiments of all organs are laid down. In addition, decapitation follows an anatomical pattern significant only after the neck has developed and the head is flexed. Nor can separation have taken place late in pregnancy, since the neck stumps are healed and the heads have disappeared, suggesting that they have been

completely resorbed. The only exception is Landau's case, where a small shrunken head remained. From its size one would estimate that separation must have occurred at 8-9 weeks' gestation.

In the case of limb amputation effects too, it is rare to find any trace of the distal portion, though shrivelled parts have been found on several occasions (Torpin and Faulkner, 1966), proving that actual amputation takes place rather than resorption *in situ*.

A consideration of the timing of the loss of the head is of vital importance in attempting to work out a possible mechanism. In the present group of acephalic fetuses, in spite of generally normal body configuration and thoracic contents, two of the authors postulated loss very early in pregnancy, for example some time in the 5 to 15 mm. stage in Case III and before the descent of the thyroid rudiment in Case IV. Klöppner allows up to the 6th month.

Benešová's suggestion that in her case it had occurred before the descent of the thyroid from the pharyngeal pouch is most unlikely. Far more significant is the atrophic and scarred state of the trachea and oesophagus, indicating a severe disturbance which may have prevented a normal thyroglossal outgrowth from developing any further, or causing it to atrophy along with the other neck organs. At a later stage in development also the cartilaginous larynx or trachea and the vertebral column would naturally resist more strongly any such influence, while the more vulnerable soft tissues above the larynx might more readily succumb. This is evident in digital amputations where cartilage also tends to be spared longer than soft tissues or bone.

The apparent absence of the adrenal glands in Case I and their small size in Cases II and IV are of little help in solving the riddle. In the anencephalic infant, the suprarenal glands are invariably small, but this is due to regression during later foetal life, since it has been demonstrated that up to about the fifth month *in utero* they develop normally (Potter, 1961). Presumably decapitation at any stage by removing the pituitary influence would lead to a similar regression, so that this factor is of little use in timing the event, except that the normal size of the suprarenals in Case III suggests that decapitation was relatively late, and possibly their absence in Case I indicates a relatively early, event.

Intrauterine decapitation has been carried out experimentally in the rabbit (Jost, 1947), with survival of the fetuses; at certain stages of gestation it leads to adrenal atrophy.

It is noteworthy in this connexion that the acephalic acardiac twins may have relatively normal suprarenals. It is possible that pituitary hormones from the other twin may be available in sufficient quantity to prevent regression.

The cause and mechanism of decapitation are a matter for debate. There is no good reason to distinguish it from other intrauterine amputation effects, except that it is usually solitary, while the majority of infants with amputation of limbs or digits show more than one lesion, often at different stages of development (Streeter, 1930; Kohler, 1962; Torpin, 1965).

Willis (1962) has reviewed current ideas on their origin, including circulatory disturbance, focal tissue deficiencies (Streeter, 1930), local vascular dysplasia (Keith, 1940), and amniotic factors. Kohler (1962) also considers this problem and its relation to the work of experimental teratologists. However, it is difficult to apply these theories and experimental results to the head and neck. More probable is a simple mechanical cause, such as Kohler (1962) maintains to be responsible for at least some of the limb defects.

Torpin's studies of the foetal membranes and placenta with relation to intrauterine amputations (1965) should bring about a reappraisal of the role of amniotic abnormalities, since his hypothesis provides a credible explanation of the mode of development of amniotic bands and their effects. He has demonstrated amniotic defects which he attributes to premature rupture of the amnion alone, followed by its separation from the intact chorion. He noted the development of strands of tissue from the bared chorionic surface and in some instances from the amnion too. In one case (Torpin and Faulkner, 1966) there was a history of threatened abortion.

The maternal histories of these decapitated fetuses and of several recorded cases of limb defects also include some abnormality during early pregnancy or evidence of premature rupture of the membranes (see Table, and Kohler, 1962). In keeping with Torpin's postulated train of events it is suggested that decapitation is a result of slow strangulation following encirclement of the neck by a ligature of amniotic or chorionic origin, ending in local scarring, separation, and finally resorption of the head.

Summary

A decapitated infant is reported. Apart from absence of the suprarenal glands, no other gross internal abnormality was present. A comparison is made between the present case and three similar

previously recorded fetuses, two of which were born alive.

Decapitation must occur during the 3rd month of gestation or later and is thought to be due to strangulation by a band of tissue of amniotic or chorionic origin.

The author would like to thank Mr. T. F. Redman for permission to publish this case and Dr. H. G. Kohler for generous bibliographic advice and help.

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