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The Absence of One Umbilical Artery in Malformed MZ Twins [1]

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SUMMARY

The authors studied the structure of the umbilical cord in a cephalothoracopagus and a holoacardius amorphus. In both cases they met with the absence of one umbilical artery. They discuss on the possible teratogenic influence of this umbilical anomaly, which appears in the acardius as a regular phenomenon, and, along with the velamentous insertion and with the vascular anastomoses bringing about the reverse blood circulation, constitutes the main cause of malformation.

In a normal umbilical cord of man we may find two umbilical arteries, and one vein of allantoic origin, whereas both omphalomesenteric arteries fade out very early. From time to time, sporadic reports appear about rare cases when one of the umbilical arteries is missing in man. In the fetuses thus affected, the incidence of further malformations is by far more frequent than in those having both umbilical arteries.

The absence of one artery has been noticed as early as 1870 by Hyrtl, who described these cases as rare anatomical curiosities. In general, we may presume, on the ground of hitherto published literature, that the absence of one artery occurs in 0.5—1% of all deliveries (Schatz, 1898; Benirschke and Driscoll, 1967; Dellenbach et al, 1968). The abortion material reveals a higher percentage, approximately 2.5% (Thomas, 1963). In a set of the perinatal departures from life, it reaches even 12% (Molz, 1965). In twins, the artery may be found missing in approximately 3.5% of cases (Benirschke and Bourne, 1960).

In the as yet outlined, relatively comprehensive sets, the data relating to MZ twins, however, are very seldom (Kaufman and Walters, 1957; Napolitani and Schreiber, 1960; Richart and Benirschke, 1963; Boronow and West, 1964; Keith et al, 1967; Slípka and Kočová, 1970), and thus we are convinced that our study of the two rare types of such twins may to some extent promote the knowledge of various problems concerning the absence of one umbilical artery.

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Own Observation

The cephalothoracopagus (Fig. 1), born in May 1965 to a 30-year-old woman in her seventh month of pregnancy as a premature child — a female — has a single head with one face (the type of *Janus monosymetros*). The neck proceeds into two, somewhat laterally connected thoraxes, with four developed upper extremities. The connection reaches down as far as the abdominal region, but two pelvises with four lower extremities are formed. No further external malformation is to be seen on the infant. There was possible, through auscultation, to ascertain a regular pulse of evidently one common heart. The child was somewhat asphyxiated, she burst out crying, and died some 90 minutes after birth.

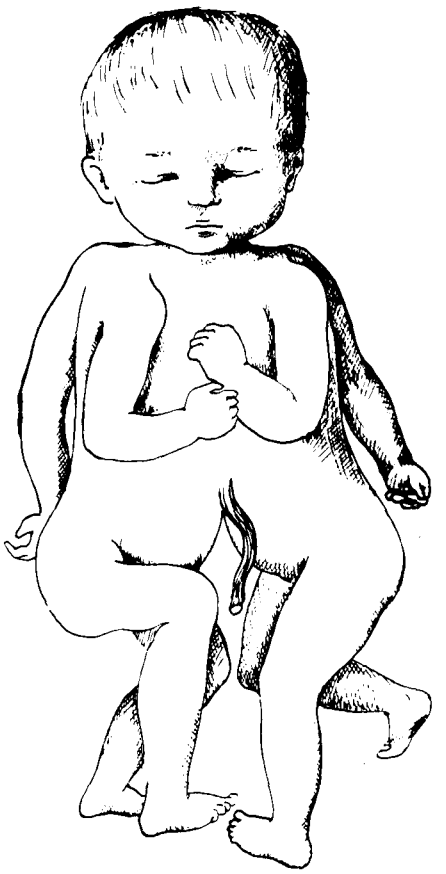

Fig. 1. Cephalothoracopagus of female sex

The body was not dissected, and is kept as a museum specimen at the Department of Gynaecology and Obstetrics of the Medical College in Baghdad. Unfortunately, the placenta has not been preserved. For our purpose, we had only a peripheral part of the umbilical cord at disposal.

On the single umbilical cord of the cephalothoracopagus (Fig. 2), a partial division into two formations of different size is marked. In the bigger part, there are the vena umbilicalis (V_1), and two umbilical arteries (A_1 and A_3). The artery A_3 is cut in a bend, so that we may see two lumina, either fused or beside each other. In the smaller part of the umbilical cord we may see the vena umbilicalis (V_2) and only one asymmetrically distended artery (A_2). In both portions of the umbilical cord we find the remnants of the allantois. In the smaller portion, the allantois is even projecting into two little branches, in both of which a lumen is preserved. This lumen is lined with several cubic cells, surrounded with very dense connective tissue, arranged typically in a concentric form.

The second specimen (Fig. 3) comes from the collections of the Department of Histology and Embryology of the Medical Faculty in Plzeň, whereto it was sent from one maternity hospital in North Bohemia in 1952. It is a specimen of the placenta of MZ twins, of the diamniotic type, with separated remnants of the membranous sac. One umbilical cord (A), attached marginally to the placenta, belongs to the normally developed female. Its preserved remainder is 20 cm long. The other umbilical cord, with a velamentous in-

sertion, measures 9 cm, and is attached to an amorphous, ball-shaped, laterally flattened formation (B), sized about $15 \times 10 \times 8$ cm, and weighing about 600 g. The surface of the acardiac offspring is covered with epidermis, provided with thin hair in the region which may be taken for the cephalic part. On the ventral side of this region, at the place where the fruit is bended, we may discern, to the right only, three little bulges which correspond

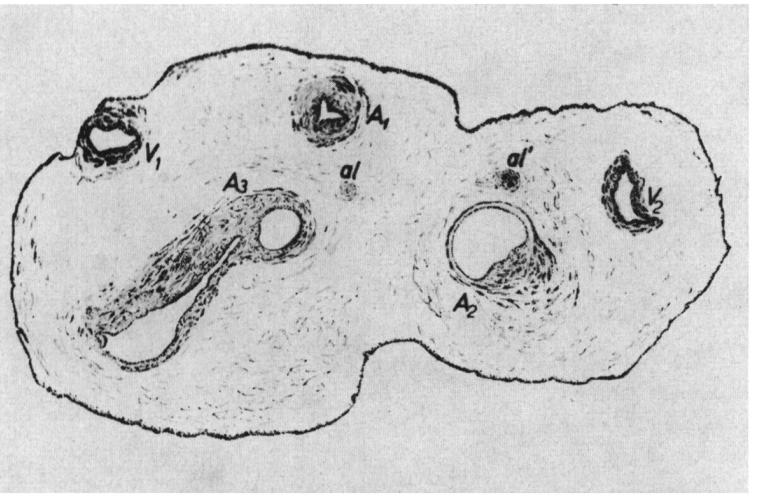

Fig. 2. Cross section of the umbilical cord of the cephalothoracopagus (A_1, A_2, A_3 = umbilical arteries; V_1, V_2 = umbilical veins; al, al' = allantois)

with the uncompletely differentiated branchial arches. The opposite side is without any formations.

The X-ray examination has not shown any sign of skeleton formation. Out of the ventral side of this part, there projects a flat umbilical cord running sideways into tiny branches in the body of the acardius — no thicker blood vessel trunks being observed anywhere.

The umbilical cord of the normal twin (Fig. 4, A) includes the usual findings inside its gelatinous connective tissue: two umbilical arteries (A_1, A_2), one vein (V), and a remnant of the allantois (al). Striking, however, is the dilatation of the vein.

In the umbilical cord of the acardius (Fig. 4, B) there are only one arteria umbilicalis (a_1), one enormously dilated vena umbilicalis (v), and, peripherally from both these vessels (cf. Thomas, 1900, and Grosser, 1927), the ductus omphaloentericus (d), plus a tiny vessel of omphalomesenteric origin, full of erythrocytes (seen only in some sections). The spacious ductus omphaloentericus has a thin wall formed of several rows of collagenous fibres and connective tissue cells to which, in the part facing the vessels, lining cubic cells with a pale-staining vacuolated cytoplasm are set. In the vicinity of the ductus we find a primitive connective tissue, characteristic for the exocoelom. This connective tissue is very thin, by which it contrasts strikingly

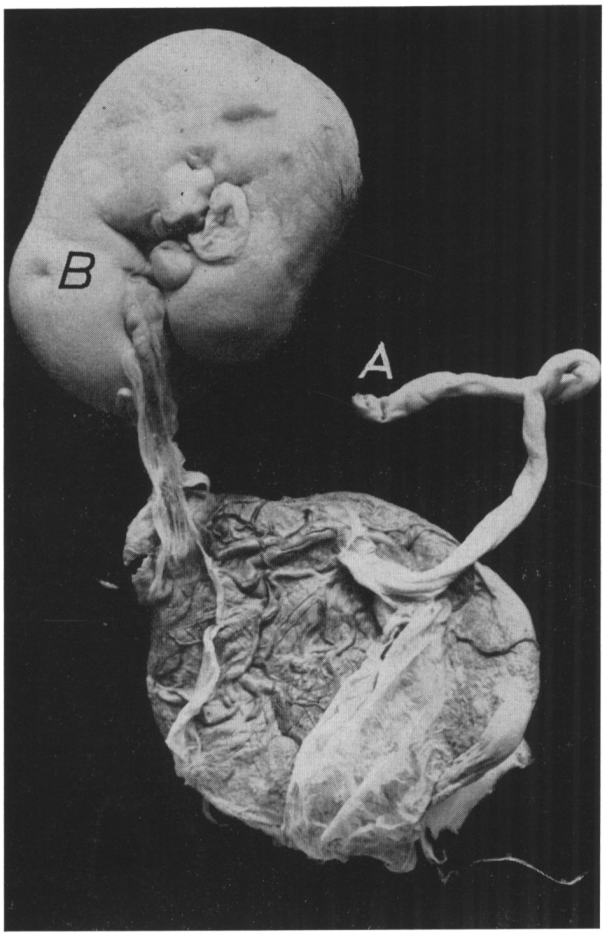

Fig. 3. Placenta with (A) umbilical cord of a normal female twin, and (B) acardius amorphus.

with the other connective tissue of the umbilical cord of the acardius, the latter being more compact than the Wharton's jelly (Zawisch, 1955). The deficiency may be caused by the decreased metabolism which affects not only the body but also the umbilical cord of the acardius.

Discussion

On the ground of the description of both cases, we may try to take up an attitude toward the question, whether the absence of one umbilical artery is the cause or the consequence of further malformations in the offspring; that is, whether there is aplasia or atrophy of the artery in question (Benirschke et al, 1864). If we accepted the theory of aplasia (Little, 1961), further malformations could be explained as its con-

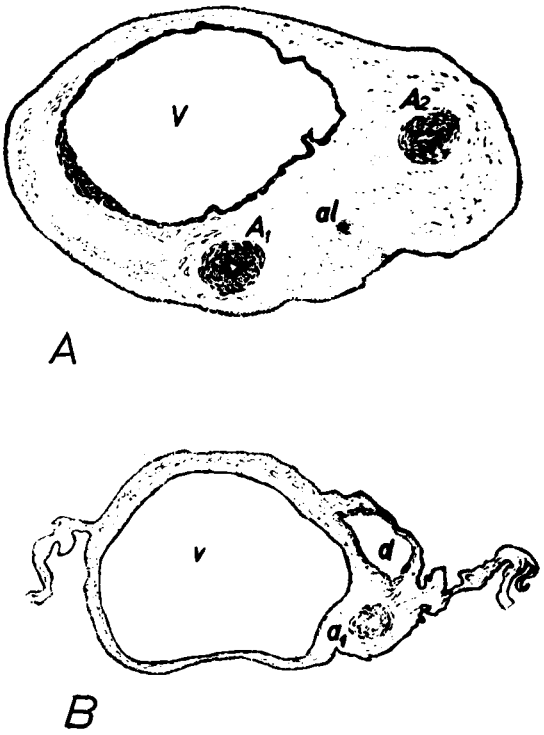

Fig. 4. Cross sections of the umbilical cord of (A) normal twin (A_1, A_2 = umbilical arteries; V = umbilical vein; al = allantois); and (B) acardius amorphus (a_1 = umbilical artery; v = umbilical vein; d = ductus omphaloentericus).

sequence (Konstantinova, 1962). The sole missing of the umbilical artery in our case of cephalothoracopagus, however, indicates clearly enough that the umbilical anomaly could not be the cause of the monstrosity, which had originated approximately three weeks earlier, before the development of umbilical arteries was completed. Hence, the absence of umbilical artery in this case is not the cause, but the sequel, of a serious developmental defect.

Quite different is the significance of the umbilical artery missing in our second case. Here it is also the question of MZ twins, even though diamniotic. The missing of the umbilical artery seems to be a phenomenon which occurs quite in the nature of a principle — in the most serious types, at least, as it is the case of the holoacardius amorphus (Napolitani and Schreiber, 1960).

The umbilical cord, itself, in the acardius, is extremely short; the placental vascular system is very limited, and the umbilical cord lies in a velamentous position, the frequency of which appears to be about 1% (Leissner et al, 1968; Purola, 1968). Apparently, the amniotic cavity, when compared to the other offspring, was much reduced. The blood vessel, both veins and arteries, anastomose (Fig. 5). The anastomoses of the acardius, however, have an immense teratogenic significance, as they change the fundamental circulatory relations in the affected fetus (Schatz, 1898;

Benninghoff and Spanner, 1929; Gedda, 1961). The oxidized blood coming from placenta through the umbilical vein to the normal fetus (A), takes in the venous blood (dashed-line arrows), led from the malformed fetus (B). The malformed fetus has not its own peripheral blood vessel. A dilation of the venous junction of both fetuses gives evidence for the cramming of blood, caused obviously both by an excessive blood flow, and by various pressure relations in both blood vessels (Dawes,

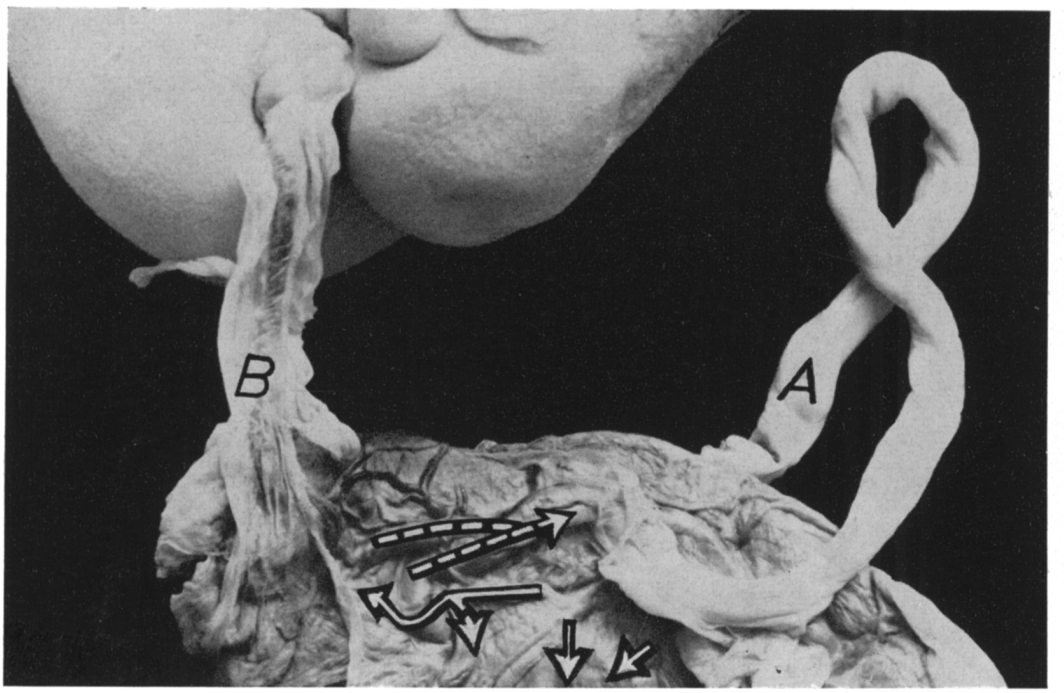

Fig. 5. Blood circulation between both twins: (A) normal twin, and (B) acardius amorphus (dashed-line arrows = direction of the venous blood; full-line arrows = direction of the arterial blood)

1962). The normal offspring gets, consequently, beside its ration of oxidized blood from the periphery, also an admixture of the venous blood from his partner.

The deoxidized blood coming from the normal fetus (A) goes away partly through the one artery in the periphery of placenta, whereas it is attached through the other to the only artery of the acardius (B), which is the sole source of nutrition for the malformed fetus (full-line arrows). Thus, it is without the least doubt that this source is quite insufficient, and that it may certainly result in an extraordinary disorder of the morphogenetic processes, and in a maximum malformation. The amorphous fetus may be considered as a parasite on the normal fetus (Schatz, 1898; Kanaev, 1959; Boronow and West, 1964), and its origin may be explained as the consequence

of the changed circulatory relations in which the absence of one umbilical artery plays certainly a prominent part.

Let us try to answer the question of formal teratogenesis of the acardius; i. e., to follow the way which resulted in the origin of the acardius. The existing theories may be divided into two groups (Dahm, 1955). On the one hand, the primary disorder of the morphogenetic proceedings is taken into consideration; on the other hand, various authors support the interpretation of a secondary disorder of the blood vessel development, which results in a circulatory insufficiency of an originally normal fetus.

On the ground of our findings, we take the liberty to pronounce a compromising hypothesis. We presume a normal, on the whole, primordium of both individuals, the MZ twins. But, as early as in the gastrular stage, the amniotic cavity in one of them is being formed less spacious. In the normal fetus, during the following expansion of the amnion, a normal strangulation of the yolk sac took place, and a normal connecting stalk, and, later on, the allantoic and, thus, umbilical blood vessels were formed. In the other fetus, the amnion expansion was less accomplished, and retarded, which caused also a delay of the strangulation of the yolk stalk in the founded umbilical cord. It becomes a mechanical obstacle of the penetrability in the umbilical blood vessels. These blood vessels reach the chorion somewhat later; in any case, after the accomplishment of the placental circulation of the normal fetus. Therefore, the attachment of the blood vessels in the malformed fetus to the existing placental vascular system of the normal fetus takes place.

Hence, we can't speak about a primary defect of the fetus, but, at most, about the primary disorder in the development of the extraembryonal structures. Neither a mere secondary influence of the circulatory disorder is here in question. The long duration of the relatively spacious yolk stalk, the velamentous position, and the persistence of the small, functioning omphaloenteric blood vessel support our hypothesis.

In conclusion, we call the attention to the fact that both studied cases belong to extreme developmental deviations; moreover, they may help in solving the question, whether the absence of the umbilical artery is the cause, or the consequence, of various associated malformations in the offsprings. From our material it may be derived that the answer cannot be quite explicit, and, consequently, this problem must be solved individually, as the case may be.

The whole problem is to be studied on a wide embryonic material or, eventually, on a comparative phylogenetic succession. Our experience, derived hitherto from individual embryos and fetuses, and, finally, from the presented twins, only confirms the general principle, stating that the absence of the umbilical artery uses to be accompanied, for the most part, with the occurrence of often multiple malformations. Hence, the absence may give a hint, already to the obstetrician, to be aware of congenital developmental defects which may often be hidden, or inconspicuous.

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RIASSUNTO

Gli autori hanno studiato la struttura del cordone ombelicale in un cefalotoracopago e in un oloacardio amorfo, notando in ambedue i casi l'assenza di una arteria ombelicale. Viene discussa la probabile influenza teratogena di questa anomalia, che si presenta nell'acardio come un fenomeno regolare, e che, insieme all'inserimento velamentoso e alle anastomosi vascolari che determinano l'inversione di circolazione, costituisce la causa principale della malformazione.

RÉSUMÉ

Les auteurs ont étudié la structure du cordon ombilical chez un céphalothoracopague et un holoacardiaque amorphe, remarquant, dans les deux cas, l'absence d'une artérie ombilicale. Ils discutent la probable influence tératogénique de cette anomalie qui se présente normalement chez l'acardiaque et qui, avec l'insertion vélamenteuse et les anastomoses vasculaires qui déterminent l'inversion de circulation, constitue la cause principale de la malformation.

ZUSAMMENFASSUNG

Verf. untersuchten die Struktur der Nabelschnur bei einem amorphen Holoakardius und bei einem Kephalothorakopagus. Bei beiden stellten sie das Fehlen der Nabelarterie fest. Sie erörtern den teratogenen Einfluss dieser Anomalie, die beim Akardius regelmässig beobachtet wird und zusammen mit der Insertio velamentosa und der Gefässanastomose, welche die Kreislaufinversion auslösen, die Hauptursache der Missbildung darstellen.

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