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Observations and Remarks on Three Cases of TRAP Sequence

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Abstract. Three cases of acardius amorphus with twin reversed arterial perfusion (TRAP) sequences are reported and the histomorphological and clinical aspects are discussed. The patho-physiology of the twin pump is briefly described. Precise prenatal assessment of the morphology of the acardius is recommended for an accurate monitoring of the perfuser in order to plan proper fetal therapy and timing for delivery.

Key words: Acardius amorphus, TRAP Sequence, Perinatal outcome.

Three cases of acardius amorphus, chorangiopagus parasiticus, with TRAP sequences are reported and histomorphological and clinical aspects are discussed.

Case Histories and Pathological Findings

1st case: First twin pregnancy, ultrasonography (*USG*) prenatal diagnosis at 31 weeks' gestation with unspecified malformation. Caesarean Section (CS) at week 33. *Placental finding:* monochorionic, bi-amniotic placenta, 750g. *Perfuser:* living, male, 1750g. Right heart failure, patent ductus arteriosus. Hypoproteinemia with oedema. Early jaundice. Surgical closure of the duct. Favourable clinical evolution. *Perfused:* 1850g acardius amorphus type E following Van Allen classification [8]. X-ray examination showed: four ribs, lumbar and sacral segments of vertebral column, ilium and ischium, femur, tibiae and fibulae. A large oedematous mass was present at the top. Peritoneum was present with ileum, appendix, colon, kidneys, ureters, bladder, adrenal glands, spi-

nal cord, gonads and male genitalia; abortive aorta and iliac arteries. The cord had 2 arteries and 2 veins.

2nd case: First twin pregnancy, without USG prenatal diagnosis. CS at week 33. *Placental finding:* monochorionic, mono-amniotic placenta, 350g. *Perfuser:* living, female, 1800g preterm idiopathic respiratory syndrome. Favourable clinical evolution. *Perfused:* 150g acardius amorphus type G according to Van Allen classification [8]. X-ray examination findings: a rudiment of ilium and femur. Oedematous mass was present. Peritoneum was absent; rudiment of colon and bladder to be seen. The cord presented 2 arteries and 1 vein, omphalocele.

3rd case: First twin pregnancy. CS at week 29. *Placental finding:* monochorionic, mono-amniotic placenta, 450g. *Perfuser:* female, 1140g. Stillborn as a result of serious subtentorial haemorrhage involving the bridge and cerebellum; lungs compatible with idiopathic respiratory distress syndrome (IRDS). Polyvisceral stasis. *Perfused:* 980g acardius amorphus type F of Van Allen classification [8]. X-ray examination revealed several ribs, portion of thoracic, lumbar and sacral segments of vertebral column, ilium and ischium, femur, tibiae and fibulae. A large oedematous mass present at the top. Absence of peritoneum. Ileum, colon, kidneys, ureters, bladder, adrenal glands, spinal cord, gonads and female genitalia present. Well developed lower aorta, iliac arteries and veins, lower vena cava. The cord showed 2 arteries and 1 vein. (Table).

DISCUSSION

In TRAP sequence, the patho-physiology of the twin pump can be briefly described as follows:

- pump stress due to perfusion of an amorphous mass which, in some cases, may present well developed organs (the greater the mass, the higher the stress);
- increased peripheral resistance due to larger placenta and to the presence of the perfused twin;
- increased total blood volume.

As a consequence of the above condition the following events take place in the perfuser: circulatory overload leading to right ventricle hypertrophy and subsequent increase in ductus arteriosus flow-rate, heart failure, liver dysfunction, congestion and hypoproteinemia which, in turn further leads to oedema reaching ascites, impaired clotting factor synthesis, impaired surfactant synthesis [5,7].

CONCLUSIONS

Considering our observations and the occurrence of all these events in our 3 cases, we would conclude as follows. The weight of the perfused twin, when related to gestational age, body growth and/or the presence of many fairly-well developed organs, is likely to be the most predictive parameter for assessing the outcome of the perfuser. Also, the presence of a greater amorphous and oedematous mass in the perfused twin seems to be correlated to a more severe outcome for the healthy twin. Accurate monitoring of

Table - Clinical and Pathological data of the 3 cases of TRAP observed

	Case 1	Case 2	Case 3
Placenta	MC-BA	MC-MA	MC-MA
Weight	750g	350g	450g
Perfused	type E	type G	type F
Weight	1850g	150g	980g
X-ray	four ribs vertebral L -S ilium ischium femur tibiae fibulae	- - ilium - femur - -	ribs vertebral T-L -S ilium ischium femur tibiae fibulae
Structures	oed. mass peritoneum ileum appendix colon kidneys ureters bladder adrenal glands spinal cord gonads male genitalia abortive aorta iliac arteries	oed. mass - - - colon - - bladder - - - - -	oed. mass - ileum - colon kidneys ureters bladder adrenal glands spinal cord gonads female genitalia lower aorta lower vena cava
Cord	2 arteries, 2 veins	2 arteries, 1 vein omphalocele	2 arteries, 1 vein

MC = monochorionic

BA = biamniotic

MA = monoamniotic

Lumbar

S = Sacral

T = Thoracic

the perfuser and precise prenatal assessment of the morphology of the acardius is recommended [2,3,6] so that proper fetal therapy and timing for delivery may be planned [1,2,5].

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