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## **Artery to Artery Twin Disruption Sequence**

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"Artery to Artery Twin Disruption Sequence" is a very rare congenital developmental malformation. This term was coined for the first time by Benirschke(l). Very few studies have concentrated their efforts to find out the incidence of this type of congenital malformation in the community. In 1989, Mohanty *et al.* reported an incidence of this malformation to be 0.01%(2).

This Artery to Artery Twin Disruption Sequence is a constellation of features due to absence of body parts because of their incomplete morphogenesis. These variably missing tissues include the head, heart, upper limbs, lungs, pancreas and upper intes-

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Received for publication: July 8, 1991; Accepted: June 8, 1994 tine. The donor twin may have an extensive cardiac load resulting in cardiomegaly with secondary liver dysfunction, hypoalbuminemia and edema. We report here a case that has most of the major abnormalities.

## **Case Report**

This baby was the fourth child of twin delivery of non-consanguineous marriage of

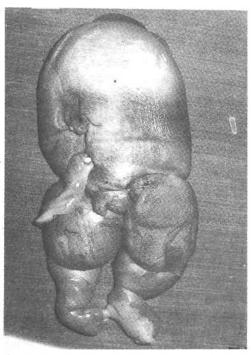


Fig. 1. Photograph showing absence of head, neck and both upper limbs, with normal trunk and both lower limbs.

Muslim parents. The 3 older sibs are normal. There was no previous family history of delivery of such malformed baby.

The pregnancy of the 24-year-old mother was uneventful till she went into labor at 30lh weeks of gestation. She had no antenatal check ups and presented at this hospital in the first stage of labor.

She delivered twin babies by breech presentation. The first baby was alive male

preterm SFD (28 weeks assessed by clinical criteria) weighing 860 g and expired after 3 days of admission in nursery. This baby had no obvious congenital malformation.

Second baby was also male still birth but congenitally malformed. The weight of baby was 1500 g with length of 23 cm. The appearance of baby was characteristic, *i.e.*, the upper half of baby (head, neck and both upper limbs) was deficient. In place of head,



Fig. 2. Photograph showing no demarcation between abdominal and thoracic cavities with visible liver and some loops of intestine.

a short, yellow colored stump was there and at upper limbs site buds were present in the form of depression. The trunk, both lower limbs with 5 toes, external genitalia with anal opening were well formed. Above and lateral to umbilical cord, a depression was present with tuft of hair (Fig. 1).

Autopsy was done, which revealed no demarcation between abdominal and thoracic cavities. Only liver with some loops of intestine, vessels were present and a myxomatous mass with depression was present in the upper part. Rest of the viscera, *i.e.*, heart, lungs, stomach, pancreas, esophagus, kidney and bladder were absent (Fig. 2).

Skiagram showed the presence of both hip bones, femurs, tibia, fibula on both sides, sacrum and vertebrae (lumbar L1-L5 and thoracic T8-T12) and 5 metatarsals and proximal phalanges on both sides were present (*Fig. 3*).

## Discussion

Benirschke reported that the great majority of monochorionic twins have conjoined placenta with vascular inter-connections. These develop on a chance basis. In artery to artery placental shunt, the tendency will be for the arterial pressure of one twin to overpower that of other usually in early morphogenesis. The defeated recipient then has reverse flow from the co-twin. This sends "Used" arterial blood from donor into iliac vessels of the recipient perfusing the lower part of the body more than the upper part. The results are a host of disruptions with deterioration of previously existing tissues as well as incomplete morpho-

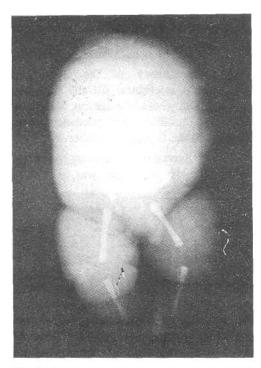


Fig. 3. Whole body skiagram showing both hip bones, femurs, tibia, fibula, and lumbothoracic vertebrae.

genesis of tissues that are in the process of formation(l).

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