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Case Report

Terata catadidyma conjoined twins: how early you can diagnose

Ashok K. Todani*, Kiranlata Todani

Department of Obstetrical Sonography, Sonography Clinic and Laparoscopy Centre, Raniganj, West Bengal, India

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***Correspondence:**
Dr. Ashok K. Todani,
E-mail: ashoktodani@gmail.com

ABSTRACT

A conjoined twin is a complex process end-result considered to be caused by either delayed splitting of the fertilized ovum or fusion of the embryonic stem cells. Disorganization of mutant genes is believed to be a cause in some cases. Conjoined twins are described as having been physically fused during pregnancy and delivery. Division after 14th day of fertilization, results in an incomplete division, because once it becomes an embryo, it is incapable of fusion. The later the division, the more severe the effect. Conjoined twins were first mentioned a long time ago when there was not much known about this. They share some organs that are vital for survival, like the heart; these twins are almost impossible to save, but there are some cases wherein there is evidence of their survival. Current technology is providing a basis for earlier diagnosis and a better prognosis. MRI and CT scan provide excellent anatomic details later in pregnancy, demonstrating organ position, shared viscera, and vascular anatomy. We present a case of live Terata Catadidyma (which refers to twins joined in the lower portion of their body, or they may appear to be two bodies on top and one body on the bottom), diagnosed at 7 weeks and 3 days by non-invasive ultrasonography, hardly reported previously so early in the literature. This reported conjoined twin were having defect in occipital region of neurocranium (forming calvarium) which should have produced encephalocele later. So, early diagnosis inflicts much less physical and psychological trauma to parents concerning viability and pregnancy outcome in long run.

Keywords: Conjoined twins, Stem cells, Siamese twins, Terata catadidyma, Zygote

INTRODUCTION

When two babies born physically connected to each other, they are identified as conjoined twins. Conjoined twins is a very rare of rare conditions characterized by fusion of separable or an inseparable part or parts of the body of genetically identical, monozygotic, monoamniotic and monochorionic twins.[1] It is still debatable whether it is due to delayed splitting of the fertilized ovum (after 13 days of ovulation) or due to fusion of embryonic stem cells that search for similar cells and attach to it as seen in case of neural tube closure.[2-4] However, gene mutation is contemplated to be the cause in some conjoined twins.[5] So, these are variations of monozygotic twins produced by incomplete dissociation of the cells of the internal cellular mass of the embryonic button, between days 9 and 13 of development. A late division of the zygote after the embryonic disc is formed, may lead to conjoined twins due to fusion. This is also called Siamese Twins. The incidence of conjoined twins is estimated to be one in 1,00,000 deliveries and the condition is more common in Africa and India. 70% of conjoined twins are females and 75% die within the first 24 hours.

A wide spectrum of conjoined defects persists as: Pyopagus: 19%, joined back-to-back, Ischiopagus:6%, joined at sacrum, Dicephalus:body one but two separate heads, Diprosopus: two faces but single head and body, Terata anadidyma: single upper body but double lower half, Terata catadidyma: twins joined in the lower portion of their body, or they may appear to be two bodies on top and one body on the bottom, Cephalopagus: 2%,

connected at head, Syncephalus: connected in the facial region, Cephalothoracopagus: connected in face and thorax, Dipygus: one upper body but two lower bodies (abdomen, pelvis, legs), Thoracopagus: 40%, joined at chest, usually share a single heart, Omphaopagus: 33%, joined near bellybutton; share liver and some GI tract, Rachipagus: back to back joined along spine above sacrum. As days pass on, the list is enlarging due to varying fusion permutations and combinations.

Methodology

After positive beta HCG urine card test, non-invasive sonography was performed trans-abdominally as a routing dating and viability assessment. Patient did not consent to transvaginal scan. There was no invasive procedure involved in it. After convincing diagnosis and consent, pregnancy was terminated and product of conception examined.

CASE REPORT

Mrs. N.G 33 years, a qualified chartered accountant, was having oligomenorrhoea and irregular periods, controlled hypothyroidism, systolic hypertension, and mild obesity. She is having one female child aged 1 year 2 months, delivered by caesarean section due to transverse lie, presented with amenorrhoea of 8 weeks and 4 days. Her urine for pregnancy test was positive on previous day at gestational age of 8 weeks and 3 days. She had no signs or symptoms of pregnancy. So, sonography scan was done for dating and to locate pregnancy site. Ultrasound examination revealed intra-uterine live pregnancy with single yolk sac (Figure 1) at 7 weeks and 3 days (Figure 2a). Nuchal translucency of one head was 1 mm (Figure 2b). Two cardiac activities (Figure 3) and (Figure 4) were noted close to each other but in two separate thoraxes (Figure 5). So, two heads, two thoraxes, but single trunk/body below was noted (Figure 6). Appearing limb buds were noted 4 at sites of upper extremity and 2 at sites of lower extremity (Figure 7) defect in calvarium was present in both heads (Figure 8a and b) was noted. Single yolk sac and single connecting stalk was identified in expelled product (Figure 9a). Bifid upper body was revealed in product of conception (Figure 9b).

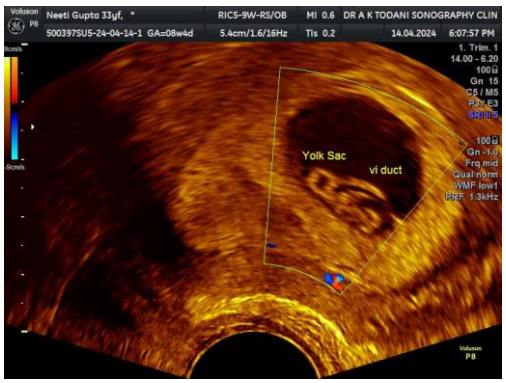

Figure 1: Single yolk sac and vitello-intestinal duct.

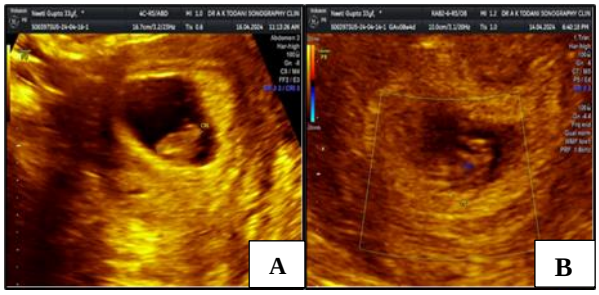

Figure 2 (A and B): CRL measurement for dating, NT was 1 mm.

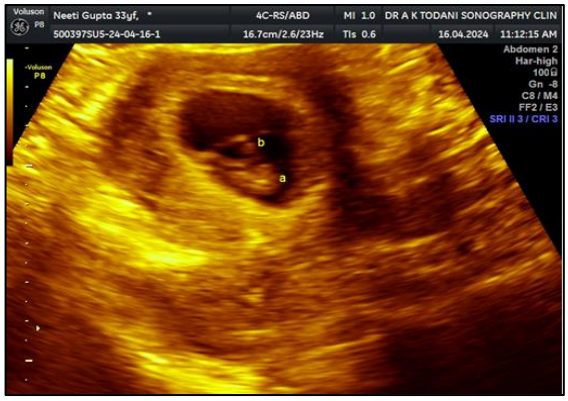

Figure 3: Two thoraxes.

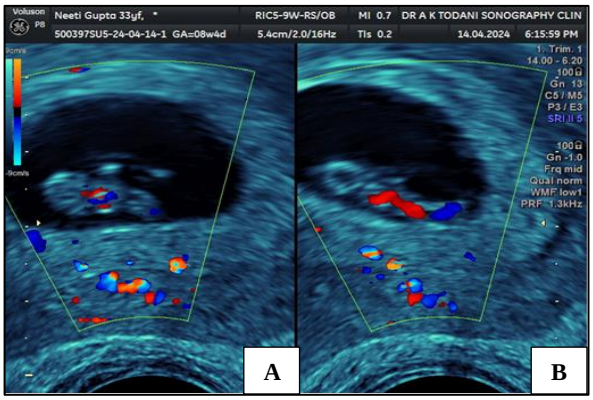

Figure 4 (A and B): Two cardiac activities very close.

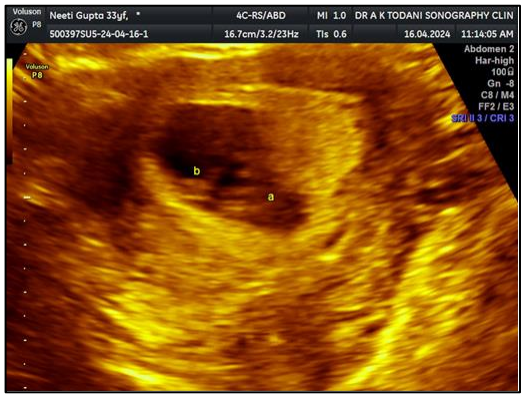

Figure 5: Two thoraxes.

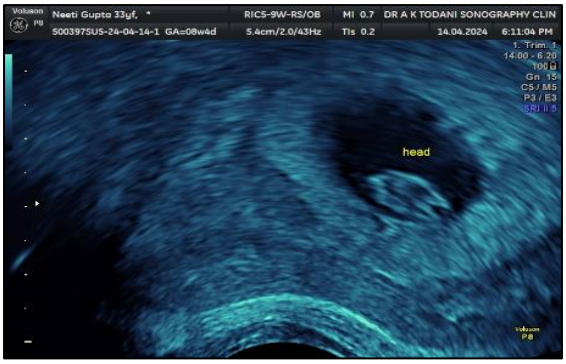

Figure 6: Foetus with bifid upper part.

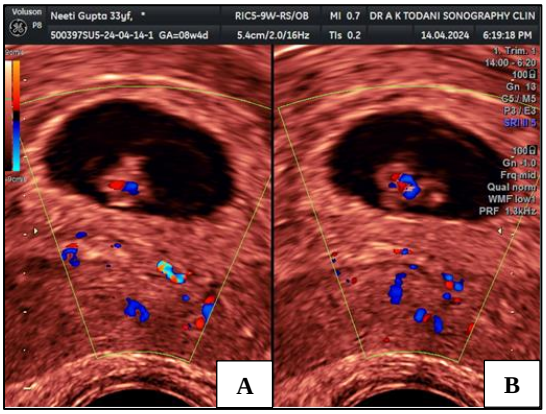

Figure 7 (A and B): 4 upper and 2 lower limb buds.

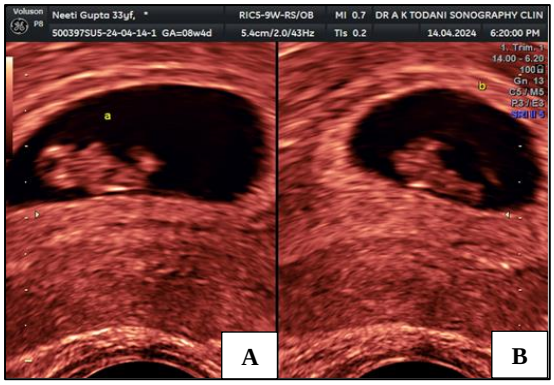

Figure 8 (A and B): Defect in calvarium.

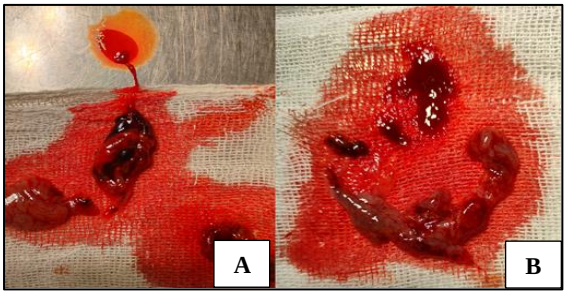

Figure 9 (A and B): Single yolk sac, cord, and placenta (left); common lower part and two upper parts put apart (right).

Considering apparent anomaly, decision was taken to terminate the pregnancy. Medical induction was done using mifegestone and misoprostol abortion kit. On expulsion of product of conception, evacuation curettage was done. Photographs of product of conception were taken. USG diagnosis criteria utilised; Demonstration of a continuous non-separated external contour, Bifid appearance of foetal pole in first trimester. Conjoined body parts, Body parts of the twins are imaged on the same level.

Outcomes

Abnormal Y-shaped distribution and configuration of foetal body was confirming Terata Catadidyma. Later in life, baby should have looked like as in diagram. Single yolk sac was present. Later in life, the baby should have been born with two heads having encephalocoele, four upper limbs but one abdomen and two lower limbs.

DISCUSSION

The earliest documented report of conjoined twins dates back to the year 945 AC from Armenia. In the Islamic history, Ibn Kathir mentioned that Hashim and Abd Shams, the sons of Abd Minaf Ibn Qusai were partially conjoined twins who were separated by their father.[6] Dicephalus twins were reported from Sardinia in 1829 and Scotland in 1940.[7] The oldest known living Dicephalus twins are Abigail and Brittany Hensel who were born in Minnesota in1990 with two heads, two arms and two legs. Dicephalus twins with 4 arms were reported from Turkey, and with 4 arms and 3 legs from Russia.[8] Conjoined twins are usually classified according to the body part at which their bodies are joined Diplopagus (pagus: fixed to) means symmetrical and well-formed twins.

Heteropagus indicates asymmetry i.e. one complete twin and a parasitic incomplete twin. Acardiac twin denotes the presence of a complete twin and another incomplete one formed of body and legs with no heart. Untill the late 1800s conjoined twins were called “monsters”.[9] The term Siamese twins come from the twin conjoined brothers Eng and Chang Bunker who were born in 1811 in Siam [now Thailand]. They were joined at the sternum by a short, flexible band of flesh and cartilage, through which their livers were connected. Triplets Tricephalus (conjoined): Extremely rare conjoining of 3 fetuses.[9] Very few confirmed cases, both human and animal, are known.[10] Prognosis: Generally unfavourable, with approximately 40% cases stillborn. Structural anomalies are frequently found such as polyhydramnios (50%), cardiac malformations, common omphalocele, and neural tube defects. Upon discovery of non-viable conjoined twins, interruption of pregnancy should therefore be recommended.

CONCLUSION

Diagnosis of conjoined twins should be made possible as early as 7 weeks gestational age, but evaluation of

common structures is not possible. Though 12 weeks scan allows clinician to assess viability, in case of non-viability, propose interruption/termination of pregnancy and avoid delayed termination hazards including psychological trauma. Let both parents and offspring not suffer from this stigma and sufferings related to conjoined twins. The new technologies such as 3D-4D sonography, 3-D printing and artificial intelligence are of utmost help in real time.

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