

A Body Stalk Anomaly with Accreta Placenta Spectrum Case: A Rare Entity

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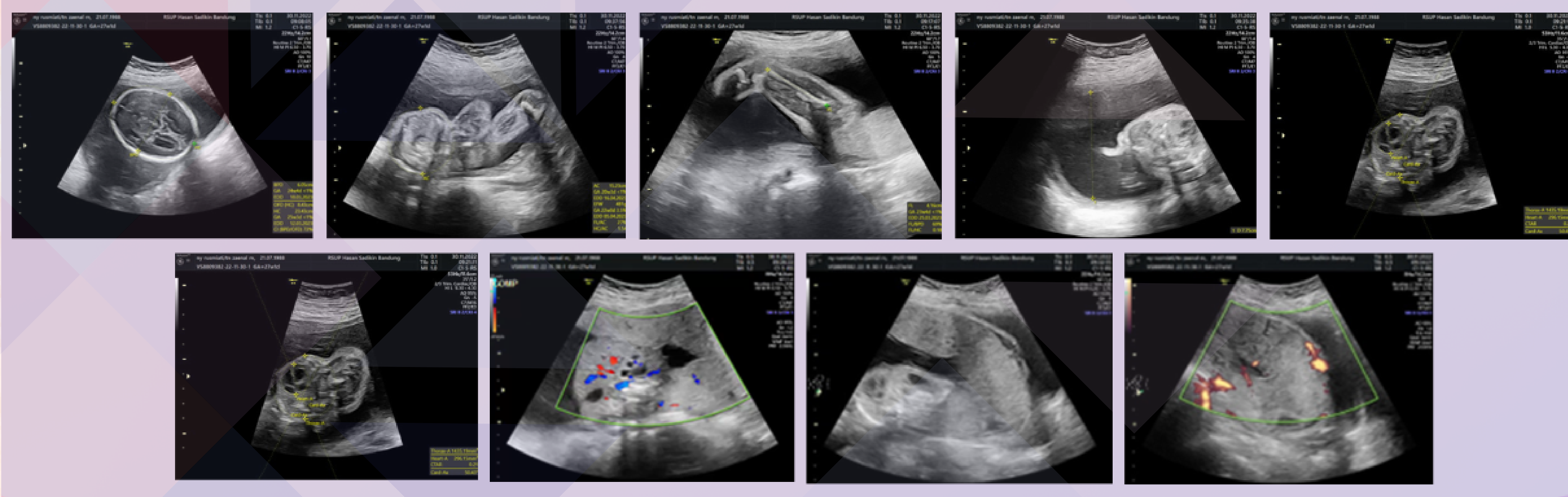
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Introduction

A **body stalk anomaly (BSA)** is a rarest sporadic condition, which occurs about **1 in 7,500 pregnancies**. It is thought to result from an early abnormal embryonic folding and defective closure of body wall in embryogenesis. While **accreta placenta spectrum (APS)** is a general term frequently applied to encompass all three of these condition (accreta, increta, and percreta).
In this case report we describe a unique pregnancy with BSA with APS.

Case Report

A **G2P1A0 34 years old woman** was referred to Maternal-Foetal Medicine Unit for further investigation of omphalocele at **29 weeks of gestation**.



An obstetric ultrasound examination: **severe abdominal wall defect, severe kyphoscoliosis, and very short umbilical cord with limb defect**. Placenta examination: **no clear zone with subplacental hypervascularization**.



Diagnosis :
A suspected BSA with APS

C-section was performed at 30 weeks of pregnancy with planning for conservative treatment for APS.

Baby : **800 gram, 25 cm length**, APGAR score of **1-1**.

Clinical examination : **a very short umbilical cord, severe abdominal wall and limb defect**.

Discussion

A body stalk anomaly has features such as **exencephaly or facial clefts or encephalocele, thoraco or abdominoschisis, and limb defects**. Possible causes of body stalk anomaly include :

- early amniotic rupture with direct mechanical pressure and amniotic bands
- vascular disruption of the early embryo
- an abnormality in the germinal disk
- defects in genes related to embryogenesis

Due to extrusion of the intra-abdominal contents, the spine and thoracic cavity do not develop symmetrically which results in severe scoliosis and abnormalities of the axial skeleton. Malrotation of the spine and incomplete closure of the pelvis can lead to malrotated limbs and/or club feet.

In this patient:

- **severe abdominal wall defect (consists of intestines, liver and stomach)**
- **very short umbilical cord**
- **limb defect**
- **severe kyphoscoliosis**.

Placenta accreta spectrum

Abnormal implantation of the placenta is believed to result from **partial or total absence of the decidua basalis and imperfect development of the fibrinoid or Nitabuch layer-the normal physiologic line of cleavage**.

At delivery, placenta fails to separate normally, leading to significantly increased maternal morbidity. **Severe hemorrhage** may necessitate cesarean hysterectomy and often massive transfusion of blood products with need for intensive care.

This patient already diagnosed as APS with lethal congenital anomaly, so this patient already chose **to terminate pregnancy**.

Conclusion

BSA itself is **a scarce entity**. Combination case with APS in this patient grant **a very poor prognosis**.

After cesarean hysterectomy, no further pre-conceptional counseling is needed in this patient.

Further counseling should include life quality improvement after surgery.