

Background

The Ultrasound work-force is experiencing inconsistencies in prenatal reporting and diagnosis within the Acrania-Exencephaly-Anencephaly Sequence (AEAS).

Anencephaly is a fatal neural tube defect characterised by the complete absence of the cerebral hemispheres including the cranial vault (Sokodziak et al., 2020). Despite being a distinct diagnosis within this spectrum of neural tube defects, Anencephaly is commonly misclassified in clinical practice as an umbrella term to describe all stages of AEAS (Santana et al., 2018). Variations in referring to specific stages of AEAS as 'Anencephaly' is widespread leading to diagnostic errors and inconsistencies in ultrasound reporting. This directly impacts decisions around management options, patient counselling and termination of pregnancy taking an emotional toll on both clinical staff and anxious patients (Salamanca et al., 1992).

Purpose

This poster aims to delineate between the distinct stages of the Acrania - Exencephaly-Anencephaly sequence and advocate the use of precise terminology within clinical practice which would improve patient understanding. This poster aims to provide a clear, evidence-based explanation of AEAS and its ultrasound appearances allowing the reader to integrate this knowledge into clinical practice. Therefore, patients will receive a precise diagnosis which will contribute to a clear understanding endorsing improved decision making. Summary of Content Illustrative examples of ultrasound cases at each stage of AEAS will be presented with recommendations of best practice including emphasis on enhancing the skills and knowledge required to make a confident diagnosis with practical recommendations for exploring standardised reporting across the field.

- Incomplete development of the cranium
- Abnormal shaped skull exposing brain tissue
- Genetic disruption of tissue that is responsible for forming bone

- Partial development of herniated brain tissue
- Exposure of brain tissue to amniotic fluid
- Mechanical and chemical damage to brain tissue

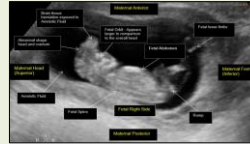
- Prolonged degradation of brain tissue
- Mostly/completely absent brain tissue and cranium
- Any remaining brain tissue is underdeveloped and non-functional

1. Acrania

Acrania is the **primary stage** in the AEAS sequence characterised by the partial absence of flat bones of the skull (Calvaria), although the brain tissue is present it is abnormally developed and exposed to amniotic fluid. This condition is commonly detected on Ultrasound imaging during the first trimester.

Key Features:

- Absence of the cranial vault (specifically the occipital, parietal and frontal bone)
- Presence of brain tissue, typically malformed and unprotected by the absence of cranial vault.
- Often progresses to Anencephaly as exposed brain degrades over time.



- Diagnosis:**
- Ultrasound (11-14 weeks)
 - Brain tissue may be seen 'free floating'
 - Absent cranial bones

3. Anencephaly

Anencephaly is the **final stage** in the AEAS sequence. The prolonged exposure of brain tissue to the amniotic fluid results in complete absence of the forebrain with little or no cerebellum. The orbits appear large in comparison to the facial features. This condition is commonly diagnosed in the second trimester.

Key Features:

- Absent calvarium, skull and scalp similar to Acrania and Exencephaly
- Absence of major portion of the brain



- Diagnosis:**
- 'Frogs eyes' appearance
 - Hyperechoic amniotic fluid due to debris
 - Polyhydramnios due to lack of fetal swallowing

2. Exencephaly

Exencephaly is the **secondary stage** or transitional stage in the AEAS sequence. The absent skull leads to direct exposure of brain tissue to the amniotic fluid causing the brain to deteriorate and appear disorganised. This condition is commonly diagnosed between the first and second trimester.

Key Features:

- Absent cranial bones
- Excessive amount of brain tissue outside the skull
- Prominent brain tissue appearing lobulated and irregular



- Diagnosis:**
- Absent skull above the orbital region
 - Disorganised brain tissue appears bulky

Summary table

Stage	Cranium	Brain tissue	Key sign	Gestation
1. Acrania	Absent/partial	Present	Beret sign	10-11 weeks
2. Exencephaly	Absent	Disorganised	Mickey mouse sign	11-15 weeks
3. Anencephaly	Absent	Mostly absent	Frogs-eyes sign	2 nd trimester

Further information

Please see the electronic stack for further information including references.

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