

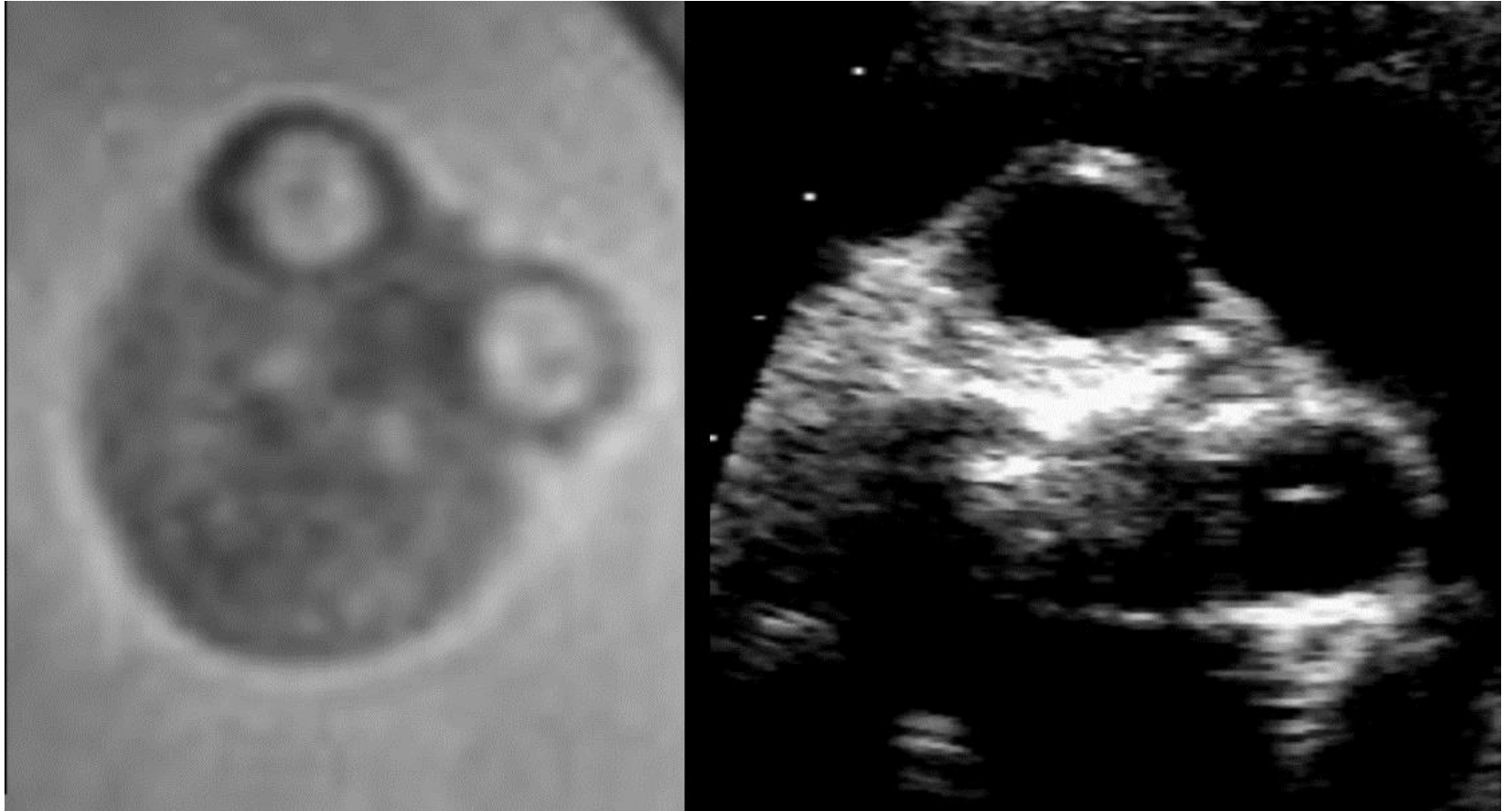
Anencephaly: pathology



Anencephaly is the absence of the cranial bones and cerebral hemispheres. It is thought to derive from the primary absence of the calvarium, with secondary destruction of the neural tissue.

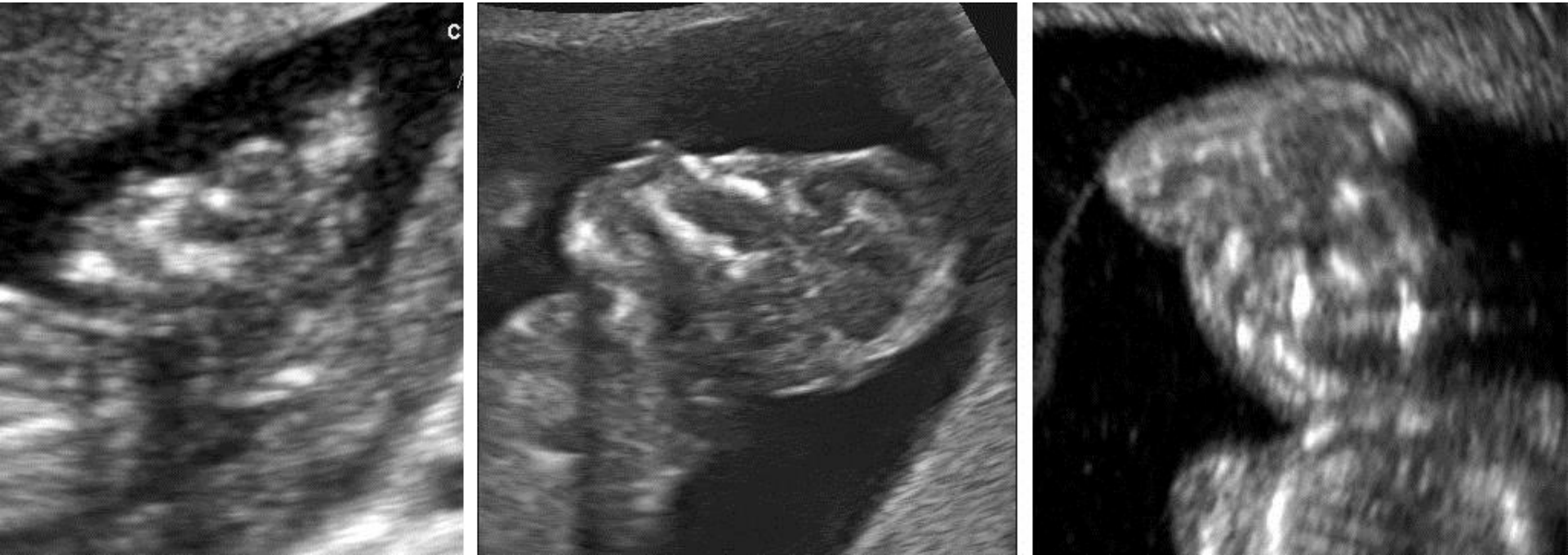
A variable amount of angiomatous stroma may be found at the skull base.

Sonography of fetal anencephaly



In the second and third trimester of gestation the main finding is the the absence of the cephalic pole above the level of the orbits.

Early sonography of anencephaly



In the early second trimester remains of the cerebral hemispheres are virtually always present, and this results in a distorted mass of variable size. The main clue to the diagnosis is the absence of the echogenic contour of the calvarium and the irregular shape of the head of the cerebral structures .

Early sonography of anencephaly: 3D

Clip 02

The contour of the cephalic pole is irregular



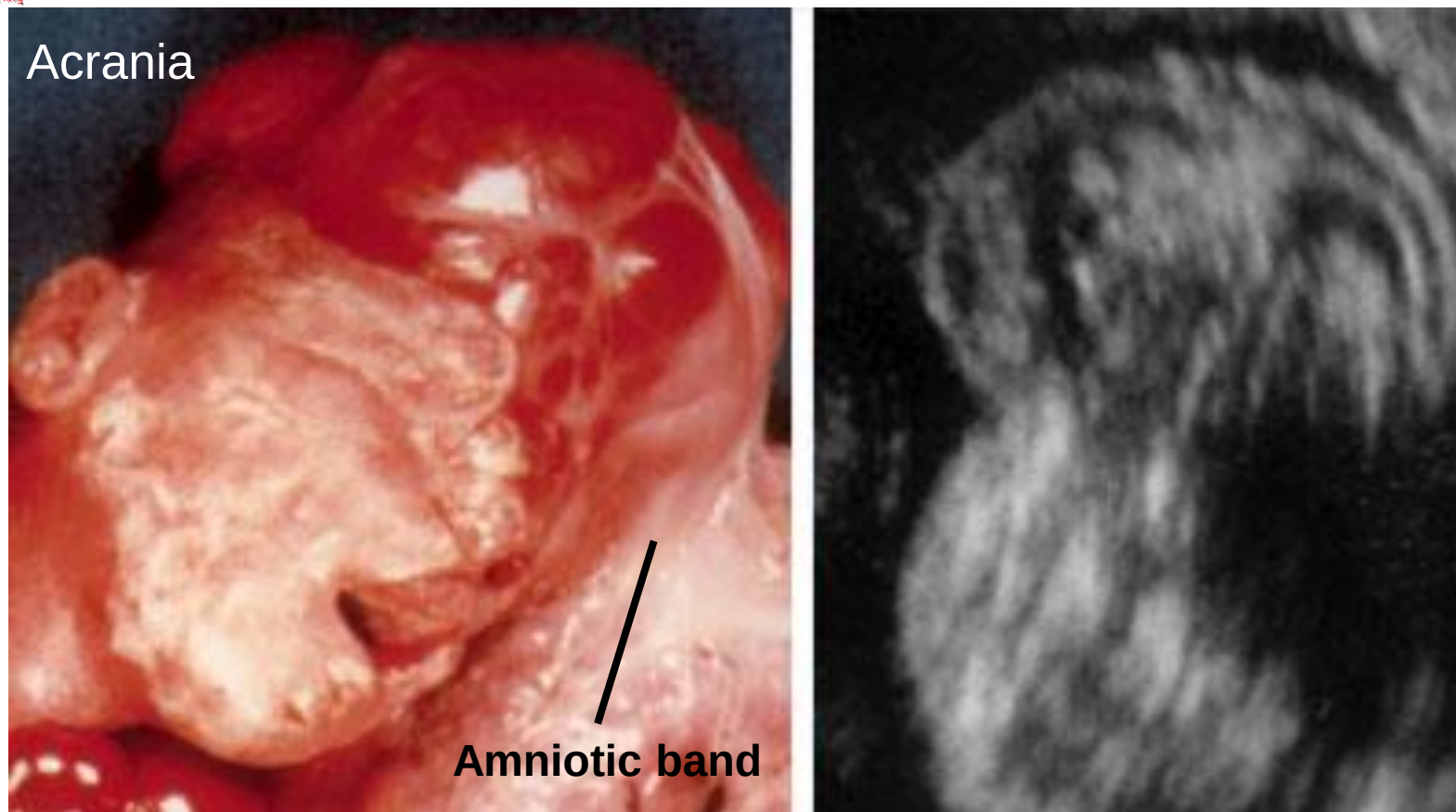
Sonography at 11-13 week: 2D

Clip 01

In real-time examinations, the soft mass of the brain floats in the amniotic cavity



Differential diagnosis: acrania



The differential diagnosis of anencephaly includes severe CNS anomalies such as severe microcephaly and gross cephaloceles. In the second trimester the absence of the calvarium in the presence of cerebral hemispheres is commonly referred to as acrania. This is frequently associated with amniotic band syndrome

Differential diagnosis: TRAP sequence

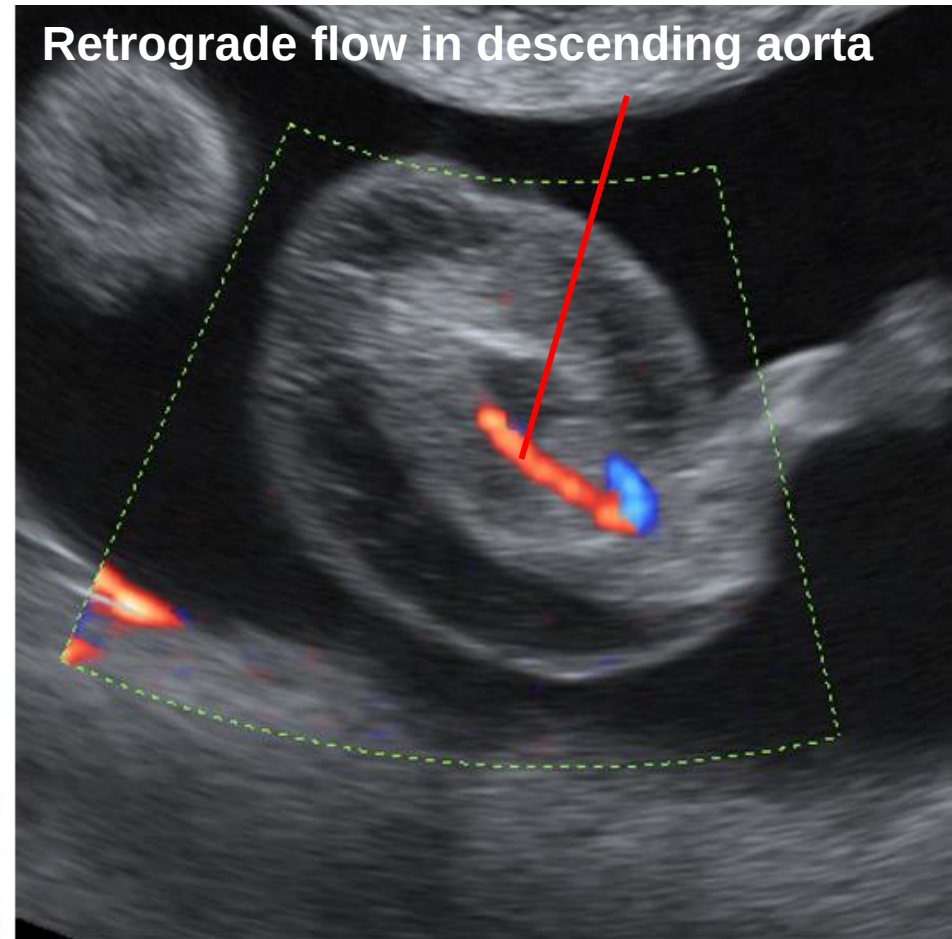
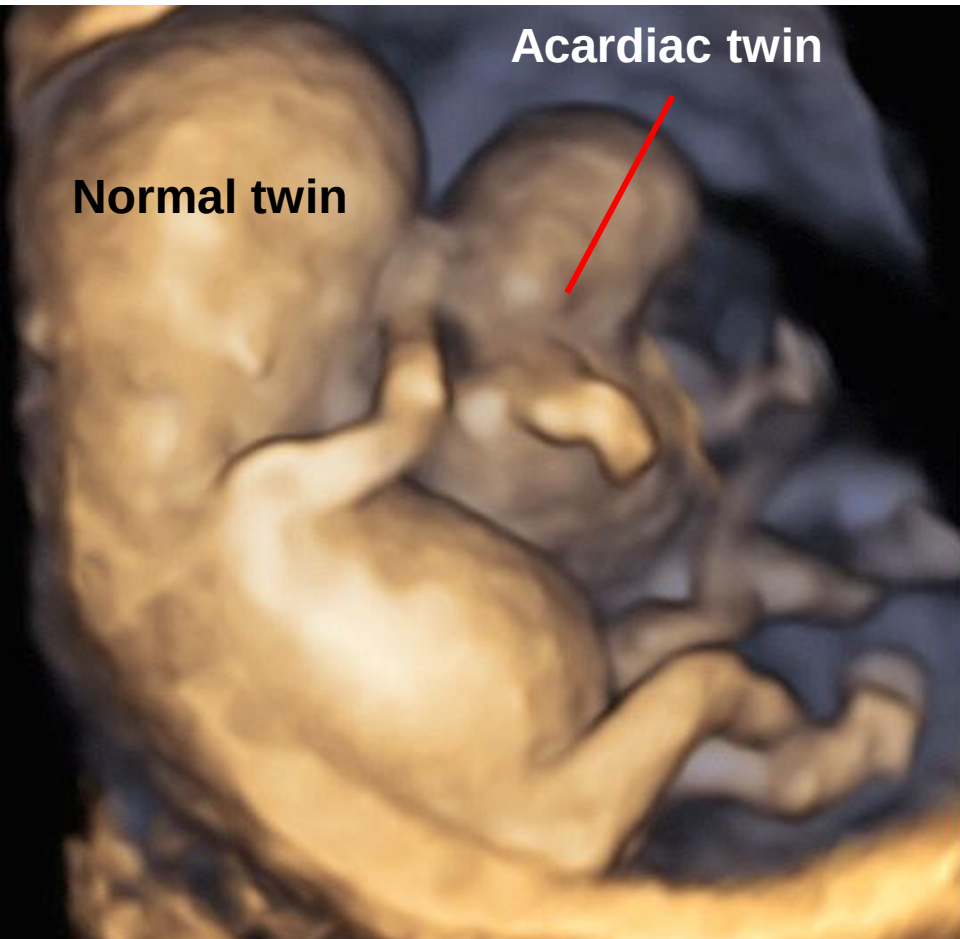


Pathologic images courtesy of www.thefetus.net

Ultrasound images by G. Pilu

In monochorionic twin pregnancies the differential diagnosis of discordant anencephaly includes the twin reversed perfusion (TRAP) sequence that usually results in an acardiac twin (acephalus-acardius). Underdevelopment of the upper trunk may lead in severe disruption of the cephalic pole with absence of the cranium.

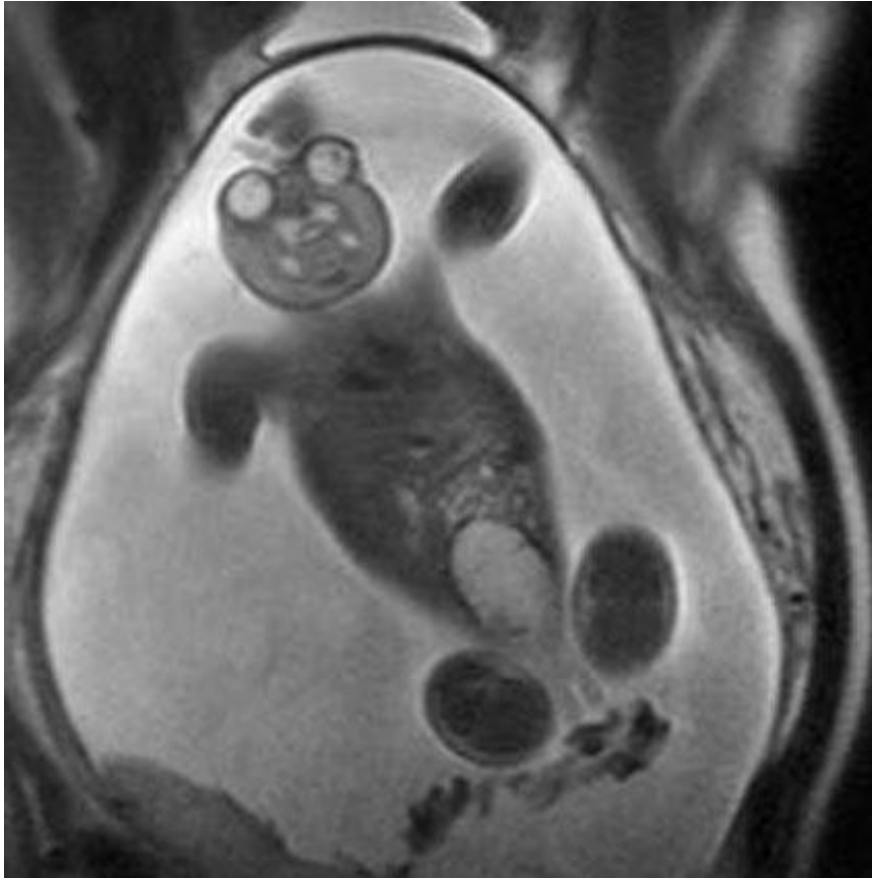
Differential diagnosis: TRAP sequence



Differentiation of anencephaly from TRAP sequence is important because the latter is associated with a much increased risk for the normal twin. The main findings of TRAP sequence include generalized underdevelopment of the trunk, subcutaneous edema and retrograde blood flow in the descending aorta.

Anencephaly: associated anomalies

craniorachisis



Polyhydramnios usually develops at the second half of the pregnancy due to failure of swallowing. Spina bifida is the most common malformation found in association. Complete opening of the spine with anencephaly is commonly referred to as craniorachisis