

Binder syndrome

Binder syndrome or maxillonasal dysplasia is a congenital condition characterised by hypoplasia of the anterior part of the maxilla and nasal complex.



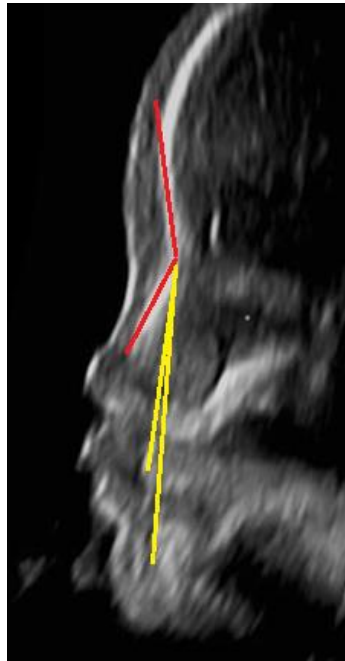
Profile views of Binder syndrome showing hypoplasia of the midface, especially in anterior-posterior direction. The protrusion of the nose and the maxilla is reduced giving the profile a concave appearance. The nose is flat (poor projection.) The length of the nasal bone is usually normal. The lips have a thick round appearance.

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Objective measurements may support the diagnosis of Binder syndrome.

An increased naso-frontal (NF) angle indicates a flat nose (red angle.)

A decreased maxillo-naso-mandible (MNM) angle indicates hypoplasia of the maxilla (yellow angle.)



Binder syndrome



Normal fetus

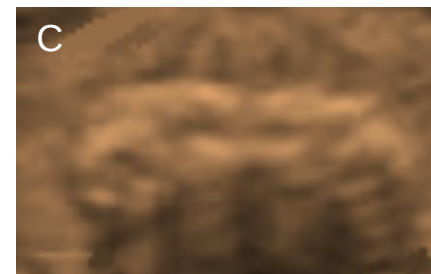
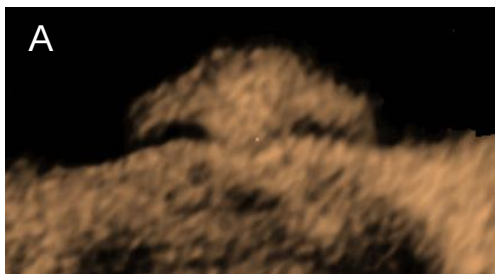
Normal values & references:

The MNM angle does not change significantly between 16 and 36 weeks' gestation, with a mean of 13.5° (the 5th and 95th centile: 10.4° and 16.9°) (E.de Jong-Pleij et al, UOG 2011;37:562)

The NF angle does not change significantly between 16 and 35 3/7 weeks' gestation, with a mean of 117° (the 10th and 90th centile: 105° and 129°) (E. de Jong-Pleij et al, UOG 2017;50:297)

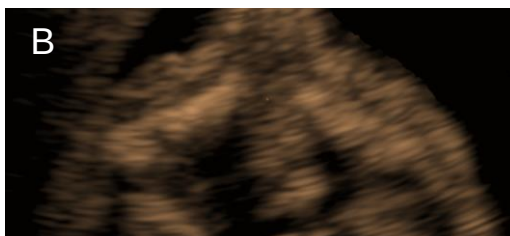
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Binder syndrome: in the 'nose-mouth view' the columella is abnormally short and the nostrils have a half-moon appearance (A). In the axial view the zygomatic arch (B) and the alveolar ridge of the maxilla (C) seems flattened.

Normal fetus



The same views in a normal fetus

Binder syndrome

Ultrasound Obstet Gynecol 2000; 16: 578–581.

The prenatal diagnosis of Binder syndrome before 24 weeks of gestation: case report

K. COOK, F. PREFUMO, F. PRESTI, T. HOMFRAY* and S. CAMPBELL



Sagittal section of the fetal face at 21 weeks' gestation demonstrating an abnormal profile with flattened nose and abnormally positioned maxilla.



3D surface rendered image of the fetal face at 21 weeks' gestation suggesting hypertelorism and flat mid-face.

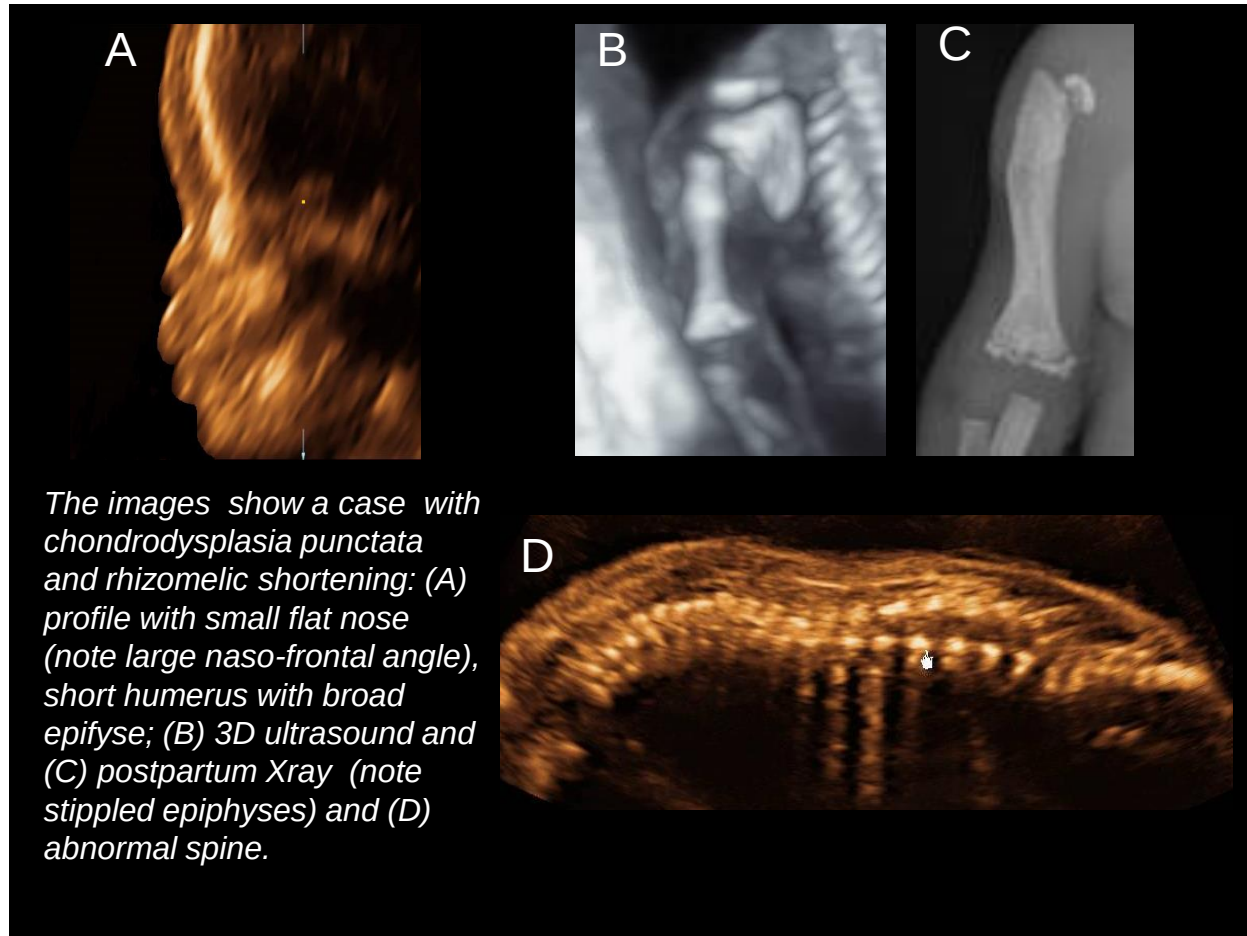
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Differential diagnosis 1

The most important condition to include in the differential diagnosis of Binder syndrome is chondrodysplasia punctata as the facial phenotype is very similar.

Chondrodysplasia punctata is a collective name of a heterogeneous group of skeletal dysplasias, having various inheritance patterns. They can be broadly divided into rhizomelic and non-rhizomelic forms. They share the features of stippled epiphyses and midfacial hypoplasia with a small nose.

The rhizomelic form can be lethal. Other anomalies encountered are spine abnormalities, ophthalmologic abnormalities (e.g. cataract), cardiac abnormalities, epiphyseal dysplasia, anomalies of the skin and hearing loss.



Prenatal diagnosis of brachytelephalangi chondrodysplasia punctata: case report

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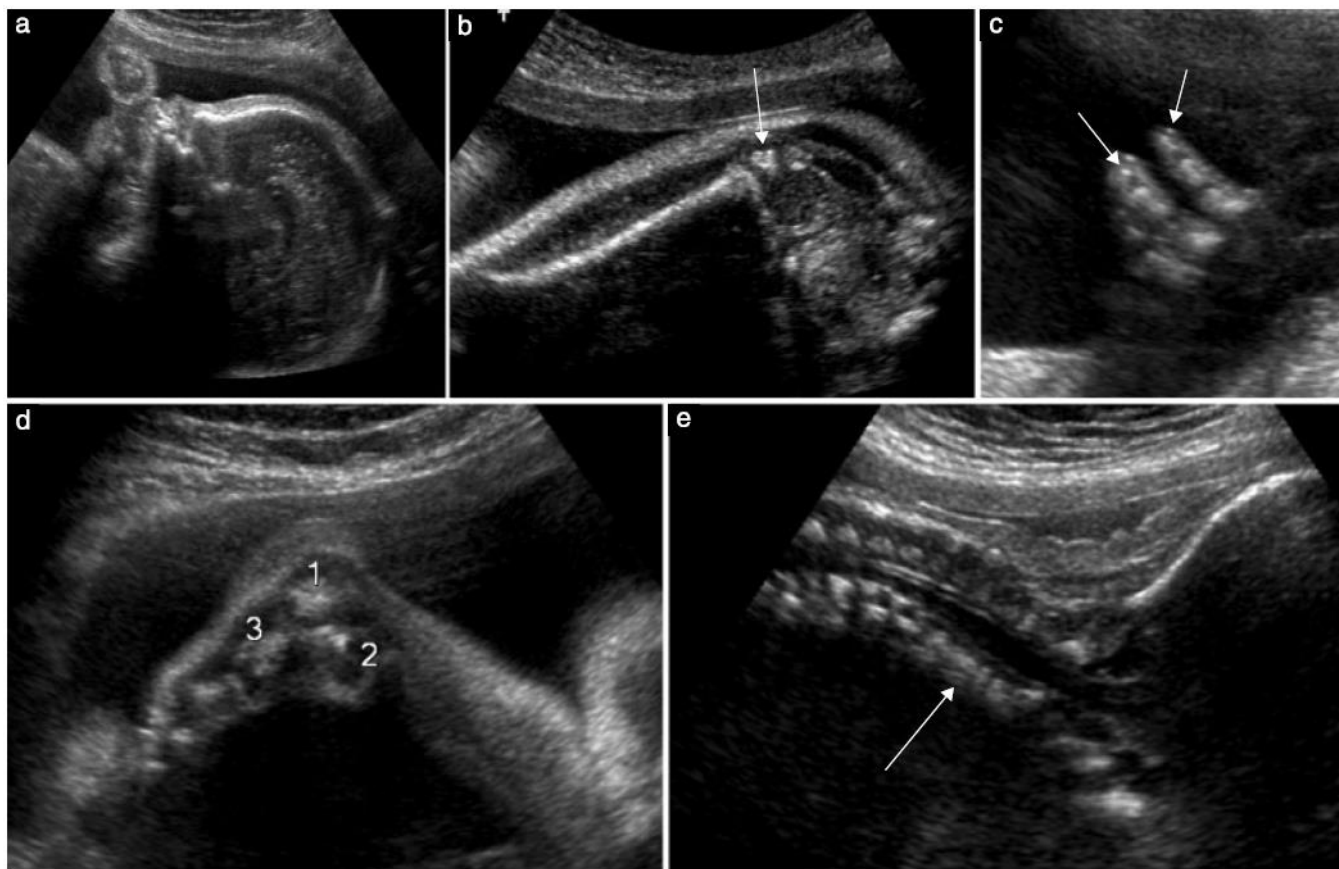


Figure 1 Prenatal ultrasound examination performed at 30 weeks' gestation. (a) Binder's dysostosis with depressed nasal bridge. (b) Proximal end of the femur with stippling close to the femoral head (arrow). (c) Second, third and fourth fingers with hypoplastic distal phalanges (arrows). (d) Lateral view of the foot. The ossification centers of the calcaneus (1) and the talus (2) are enlarged owing to stippled calcification. The ossification center of the cuboid bone (3) is visible prematurely. (e) Lateral view of the cervical spine. Diffuse stippling of the vertebral bodies (arrow) is responsible for their blurred appearance.

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Differential diagnosis 2

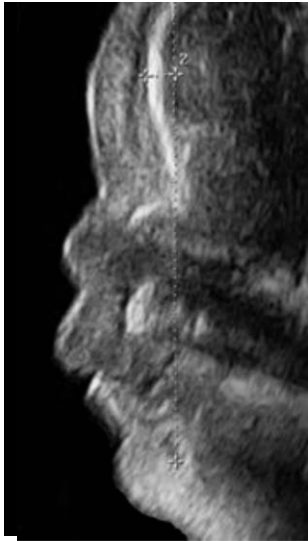
Next to chondrodysplasia punctata there are several syndromes with flat noses and midfacial hypoplasia. Some are summarised in the table below.

Condition	The face	Other anomalies	Inheritance
Wolf-Hirschhorn (4p deletion)	Broad, flat nasal bridge (Greek warrior helmet) High forehead Hypertelorism Short philtrum Small downturned mouth Sometimes: micrognathia, poorly formed ears and clefts	Microcephaly Seizures Delayed growth and development Intellectual disability Hypotonia Scoliosis and kyphosis Sometimes: abnormalities of the eyes, heart, genitourinary tract, and brain	Most cases are not inherited, but are the result from a chromosomal deletion or a ring chromosome 4 formation during reproduction. Sometimes a parent carries a balanced translocation
Keutel syndrome	Depressed nasal bridge Small alae nasi Hypoplasia of the mid-face Sloping forehead Retrognathia	Diffuse cartilage calcification Pulmonary stenosis, Hearing loss Short distal phalanges Hypotonia Mild mental retardation	Autosomal recessive
Robinow syndrome	Depressed nasal bridge Hypoplasia of the mid-face Bossing forehead Hypertelorism	Mesomelic limb shortening Hypoplastic external genitalia in males Orofacial abnormalities Renal anomalies Vertebral anomalies	There are both dominant and recessive forms of Robinow syndrome
Aarskog syndrome	Small nose with broad nasal bridge Anteverted nostrils Long philtrum Maxillary hypoplasia Hypertelorism Widow's peak hairline Ear abnormalities	Genital abnormalities Brachydactyly / clinodactyly Pectus excavatum Spina bifida occulta Mild learning difficulty and/or behavioral disorders Sometimes: scoliosis, cleft palate and eye abnormalities	X linked recessive

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Differential diagnosis 3

Syndromes with a small noses (generally small, not especially flat) are numerous. Some conditions are shown on the images below.



Pallister Killian syndrome
Note the generally small nose. The naso-frontal angle is not increased and the maxilla is not hypoplastic. There is a long philtrum and prefrontal edema.



Stickler syndrome
Note the generally small nose. The naso-frontal angle is not increased and the maxilla is not hypoplastic.



Thanatophoric dysplasia type 2
Note the small nose with a depressed nasal bridge. There is also frontal bossing. The maxilla is not hypoplastic



Campomelic dysplasia
Note the generally small nose. The philtrum is long. There is no hypoplasia of the maxilla.

It should be remembered that the shape of the nose is influenced by racial differences and familial traits, especially later in pregnancy.