# Congenital Epulis: a clinical case presentation

**Kristie Sweeney**[1]
AMS, BAppSci (Medical Imaging), Grad. Dip. Med. Sonography

**Jacqueline Spurway**[1]
AMS, MHlthSc (Medical Sonography)

**Brendan Mein**[2]
AMS, BSc. Grad. Dip. Med. Sonography

**Robert Magotti**[2]
FRANZCOG, DDU

**Ron Benzie**[2]
MBChB, FRCOG, FRCSC, FRANZCOG, ARDMS

**Daniel Challis**[3]
FRANZCOG, DDU, CMFM

**Guy Henry**[4]
MBBS, FRACS (Paed, Gen)

[1]Orange Health Service
Orange
New South Wales
Australia

[2]Perinatal Ultrasound
Nepean Hospital
Penrith
New South Wales
Australia

[3]Royal Hospital for Women
Sydney
New South Wales
Australia

[4]Sydney Children's Hospital
Sydney
New South Wales
Australia

Correspondence to email
Kristie.Sweeney@health.nsw.gov.au

**Abstract**

Congenital Epulis (CE) is a rare, benign tumour of the mucosa of the mouth in a neonate. It presents as an intraoral tumour and is rarely diagnosed prenatally. Complications include neonatal airway compromise, difficulty feeding and aesthetic considerations. Ultrasound is useful in aiding decisions regarding site, age, method of delivery and preparing parents and staff for the appearances of the tumour at birth. We present a case where CE was identified at 35 weeks gestational age during a routine third trimester prenatal ultrasound.

The patient was scanned at a rural centre, referred to a tertiary institution for follow up and delivered at a specialist perinatal surgical centre, in preparation for neonatal surgery. The outcome was excellent and this case is a good example of multi-centre cooperation.

*Keywords*: congenital epulis, congenital granular cell tumour, Neumann's tumour, prenatal diagnosis.

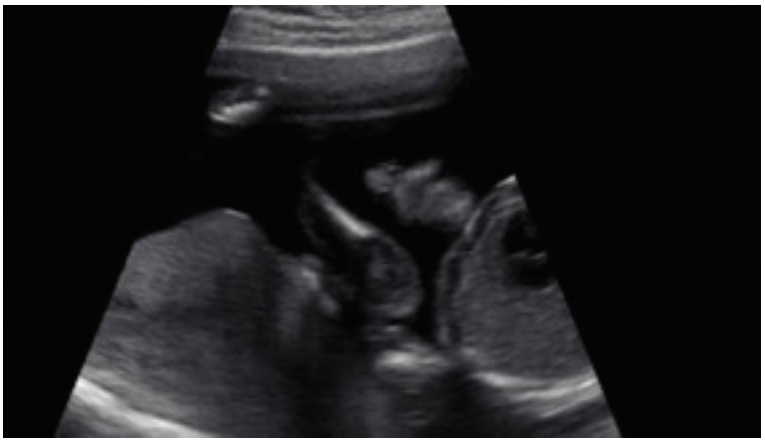

**Figure 1:** 20 week coronal face.

## Introduction

Congenital Epulis (CE) is a rare, benign tumour of the mucosa of the mouth seen only in the neonate.[1–13] It was first described by Neumann in 1871[1–4,6,9] and as such it is also called Neumann's tumour or a congenital gingival granular cell tumour.[5–9,12,13,14]

CE is commonly diagnosed postnatally. Approximately 200 cases have been reported in the literature[1–3,9]; with only 9 cases diagnosed prenatally.[1]

CE most often presents as a single intraoral soft tissue mass of variable size,[1,9,13] usually arising from the maxillary alveolar ridge.[1,8–13] However, it has also been reported arising from the tongue and mandible[2–4,6–9,12,13] and as multiple lesions in 10% of cases.[2–4,7] Its size is variable, ranging from a few millimeters to 9cm.[1,3] It is more common in females, predilection rate reported at 8:1–10:1.[6–10,12]

Spontaneous regression has been reported,[9,12,13] but surgical excision is the most common treatment. CE has not been reported to reoccur[2,4,5] nor has malignant epulis ever been reported.[2,4–8] According to Nyberg, *et al*,[11] fetal facial tumours are not generally associated with any chromosomal abnormalities, although CE has been diagnosed in a female XXX fetus.

## Case report

### *Orange Health Service*

A 37-year-old primigravida presented for a routine morphological assessment at a gestational age of 20w 4d calculated from a known LMP. A privately performed NT ultrasound returned a low risk result and the pregnancy had so far been uneventful. At the 20 week ultrasound the fetal biometry was concordant with known dates and the morphological assessment was unremarkable. The placenta was posterior and low lying and a repeat scan was recommended in the third trimester.

The patient returned at 34w 4d gestation at which time the fetal presentation was flexed breech and the placenta was well clear of the internal os. There had been good interval growth of all fetal biometry and the estimated fetal

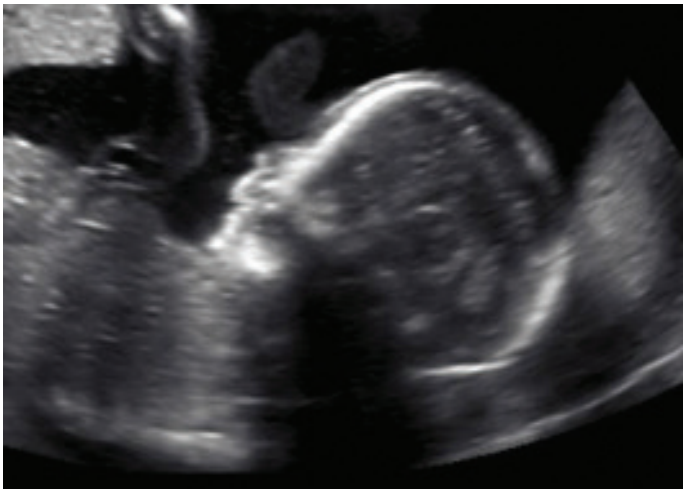

**Figure 2:** 20 week face profile.

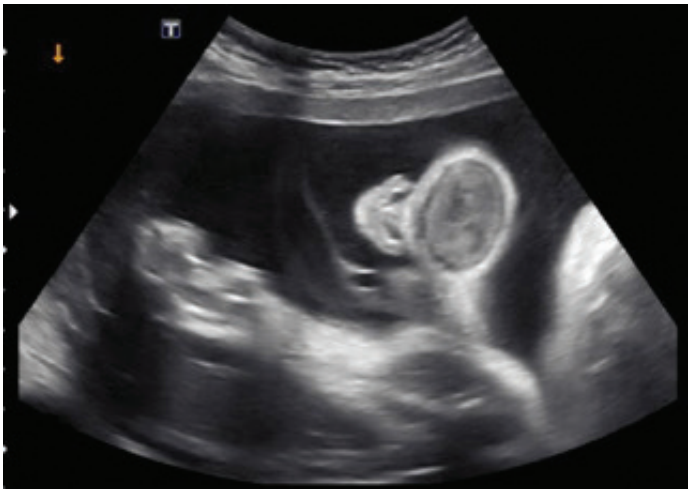

**Figure 3:** 35 week coronal face.

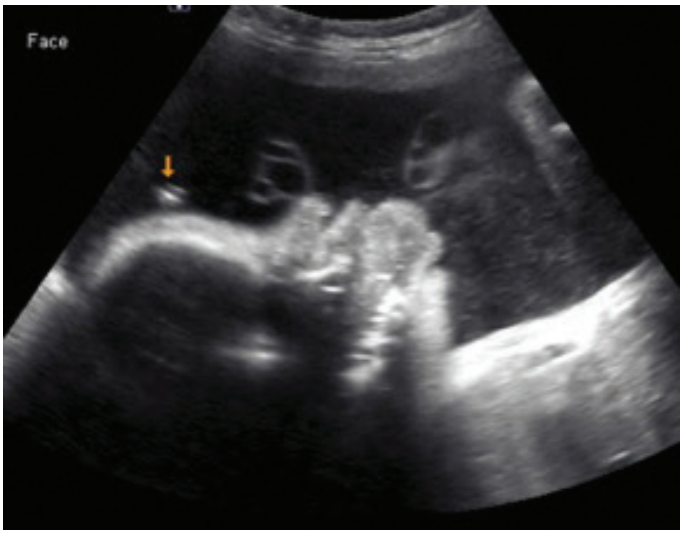


**Figure 4:** 35 week face profile.

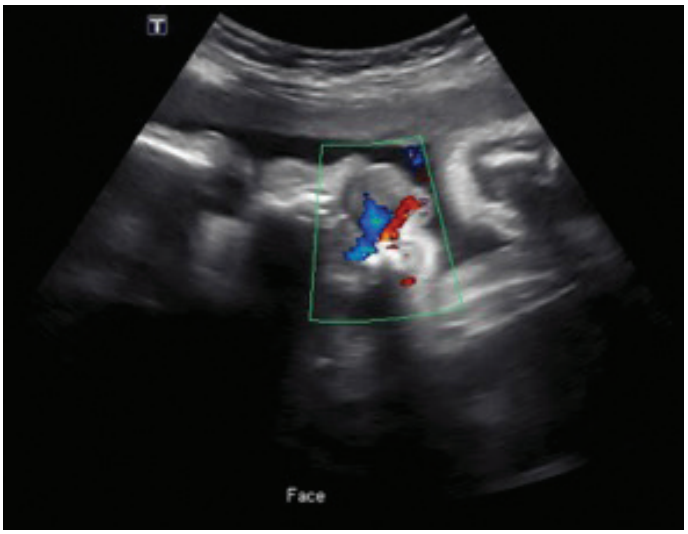


**Figure 5:** 35 week face profile CFD.

weight was 2599 g (62nd percentile). The AFI and umbilical arterial Doppler measurements were within normal range for gestational age. A homogenous mass was identified protruding from the fetal mouth. There was obvious vascularisation of the mass arising from its inferior aspect. The mass could not be differentiated from the tongue and posterior extension into the pharynx was not excluded. The patient was referred to Nepean Hospital, a tertiary facility, for review.

### *Nepean Perinatal Ultrasound*

A review scan was performed at 35w 2d. A well grown female fetus, 2428 g (26th centile), was seen, with a normal AFI and stomach bubble.

A solid mass was identified protruding through the fetal mouth, separate from the tongue, measuring 26 x 19 x 21 mm. The mass appeared to originate from the mandible and had a vascular pedicle. The fetus appeared unable to close its mouth. The nasal passages were seen to allow bidirectional flow of amniotic fluid on Colour Flow Doppler (CFD) mapping implying uninterrupted nasal breathing and hence obviating the possible need for an EXIT procedure at the time of delivery.

The remainder of the fetal anatomy was normal. These findings were consistent with congenital epulis or oral teratoma. The patient and family were counselled and were referred to the Royal Women's Hospital for delivery and definitive management as paediatric surgical services were available on site.

### *Royal Hospital for Women and Sydney Children's Hospital*

The patient was reviewed at the Maternal Fetal Medicine Department and discussed in the multidisciplinary team meeting involving paediatric surgery and neonatology, as well as fetal medicine specialists. The provisional diagnosis for the lesion was CE, although teratoma was not excluded.

The fetus was delivered by caesarean section at 39 weeks at the Royal Hospital for Women and Children, with paediatric surgical attendance. Birth weight was 3570 grams (71st centile). No resuscitation was required with APGAR of 9 at 1 and 5 minutes. The baby was admitted to NICU. At day 2, the epulis was surgically excised under general anaesthesia without complications and the baby was discharged on day 3. The baby continues to do well.

### *Final Pathology Report (Summary)*

Specimen was a nodular lesion covered by pale shiny mucosa measuring 25 x 15 x10 mm and had an area of ulceration 15 x 5 mm.

Sections show a polypoid mass lined by keratinised stratified squamous epithelium consistent with gingival

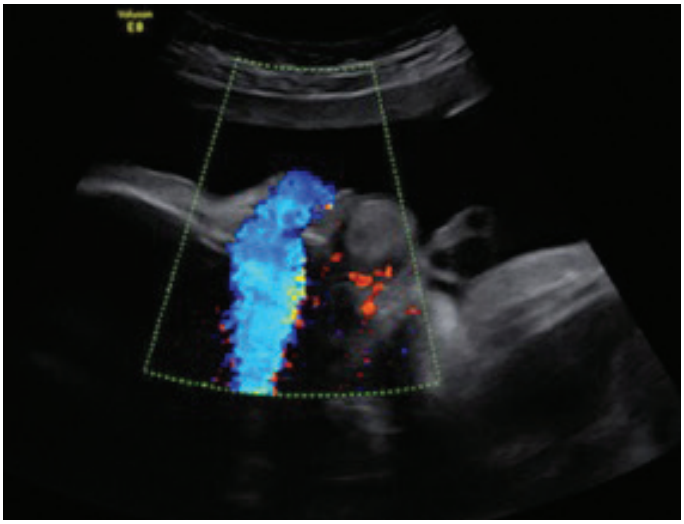

**Figure 6:** CFD showing bidirectional flow of amniotic fluid in nasal passages.

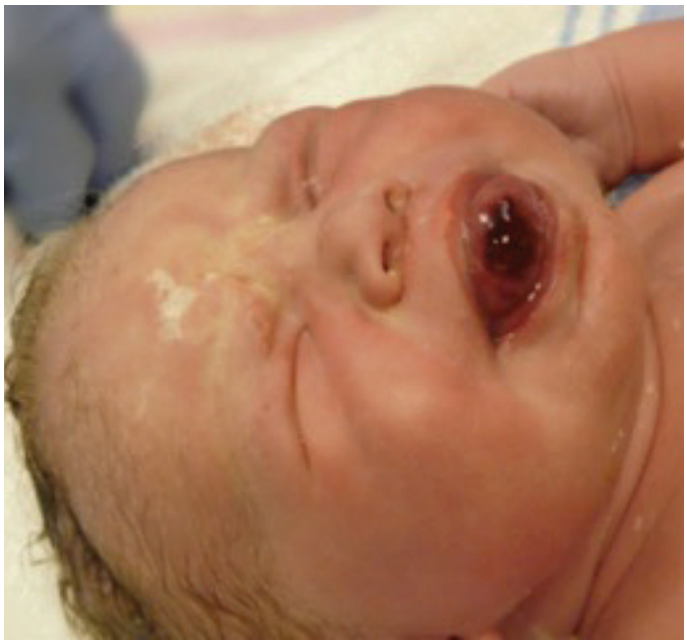

**Figure 8:** CE as seen at birth.

mucosa. The sheets of lesional cells were separated by a rich vascular network which includes admixed perivascular and interstitial lymphocytes. There were areas of dystrophic calcification. Occasional eosinophils were present among the lymphocytes. There was no necrosis and mitoses were not identified. Focal inflammation of the mucous was present associated with submucosal clefting and acute inflammatory infiltrate. The immunohistological stains showed the lesional cells were negative for S100 and SMA. The lesion did extend to the diathermied margin of the resection.

Diagnosis – Mouth Congenital Epulis.

## Discussion

### *Etiology and Pathogenesis*

The exact cause of CE remains unknown and controversial.[3,12,13] There have been several theories, but none have been proven. Initially CE was thought to be a form of ondontogenic dysgenesis,[2] however, there have been no reports of any other

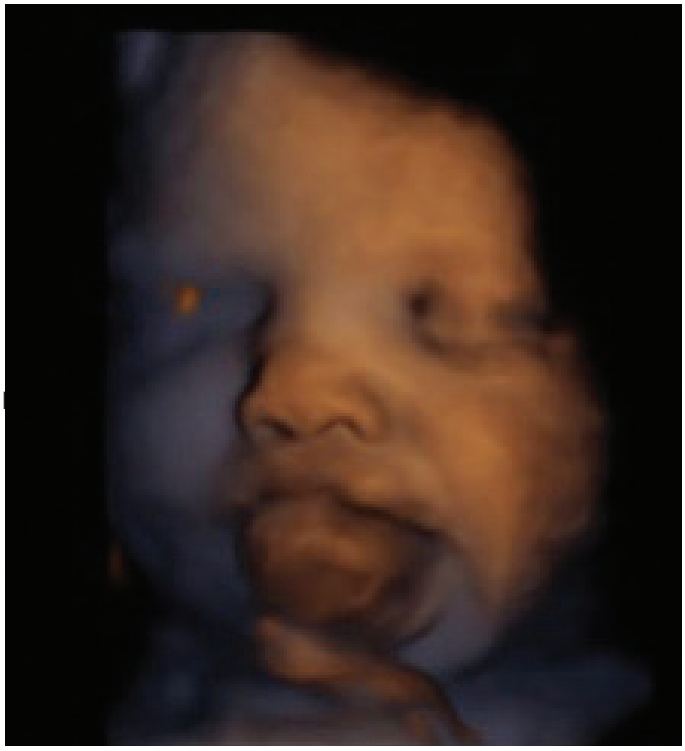

**Figure 7:** 3D face.

**Figure 9:** Complete recovery several months after surgery to remove CE

dental abnormalities associated with CE, except for rare accounts of hypoplastic or absent teeth.[3,4,9] Due to the high female to male ratio, an intrauterine hormonal stimulus was proposed as a cause of CE, but was disproved as there are no hormonal receptors in the tumour.[7,12,13] CE is now thought to be a reactive or degenerative lesion with a mesenchymal origin.[2–4] CE has never been observed to increase in volume after birth,[2] or show malignant conversion[2,4–8] or recur after excision.[7–11,13] CE is not generally associated with any other congenital abnormality.[1,8,9]

### *Prenatal diagnosis by US*

Ultrasound has only rarely diagnosed CE prenatally.[1,3,11] CE has been discovered coincidentally in third trimester scans, but has never been reported before the 25th week of gestation.[3,6,7] A number of these patients had unremarkable scans previously.[1,3,8,11.] Wittebole, *et al.*[13] report a patient diagnosed with CE at 27 weeks, which increased in size from 12 x 11 mm to 81 x 40 mm in the 37th week. These observations may suggest that CE has an accelerated development and growth pattern in the later stages of pregnancy.[1,9,11]

Prenatal diagnosis is important for several reasons:

1 Parental counselling – a neonate with a CE can be an alarming sight for parents and healthcare professionals.[8] Counselling regarding the probable size of the tumour is useful in preparing parents. 3D Ultrasound could also improve parental understanding.[5]
2 Airway and fetal swallowing obstruction – monitoring of the size of a CE is beneficial, as larger lesions are more likely to obstruct the airways at birth.[1,4–6] This has also been observed in multiple epuli.[1,5] Assessment of fetal swallowing can rule out airway obstruction,[9]which can cause polyhydramnios[1–3]
3 Difficulty feeding – a CE could make neonatal feeding difficult or impossible.[3–9,13]
4 Delivery options – if fetal airway obstruction or distress at birth is a possibility, caesarean section with ex- utero intrapartum therapy (EXIT) procedure can be considered.[1,5,9] An EXIT procedure is a specialised procedure that maintains utero-placental blood flow for fetal oxygenation after the uterus has been opened. The fetal head is delivered while another team secures its airway. This allows time for the paediatric team to work on establishing the airway, and then complete delivery of fetus and placenta once the airway is secured[14.]

It is not possible to positively confirm the diagnosis of congenital epulis by ultrasound alone. Findings are non- specific and there are several differential diagnoses. Magnetic Resonance Imaging has been used to confirm the gingival origin of tumours and also to exclude extension of the mass,[1,5,7] however, CE must be confirmed by histopathology.[1,8,12]

### *Differential diagnosis*

Differential diagnosis can include:

- Oropharyngeal teratomas[1,8,9]
- Haemangioma[1,3–9,13]
- Anterior encephalocele[1,5,8]
- Cystic lymphangioma[4,8,12]
- Digestive or tongue duplications[1,3]
- Fibroma[9,12,13]
- Rhabdomyoma and rhabdomyosarcoma[9,12,13]
- Schwannoma[3,13]

### *Treatment*

Although spontaneous regression has been reported,[5–9,12,13] it was only seen in small tumours.[1,8,9] Surgery is the treatment of choice. Surgical excision and excision by laser with local and/or general anaesthesia has been described.[1–3,12,13] Radical resection is unnecessary as the tumour has not been known to recur even when clear margins were not achieved.[1,3–5,10]

## Conclusion

Congenital epulis is a rare, benign tumour of unknown origin, which usually arises from the maxillary mucosa of neonates. Typically, CE presents as a single tumour. It does not increase in size after birth, nor recur after excision and it has never been reported as malignant. It appears to develop and grow in the late 2nd and 3rd trimesters.

CE, when diagnosed prenatally and confirmed histologically, has an excellent outcome. US can establish airway patency with Colour Doppler and diagnose polyhydramnios due to swallowing difficulties. In case of possible airway patency problems, arrangements for an EXIT procedure at delivery need to be organised.

The management of this case shows how good collaboration between hospitals with different role delineations can result in very good outcomes for patients.

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