# Tracheal bronchus in a 6-month-old infant identified by CT with three-dimensional airway reconstruction

A 6 month old infant presented with a history of cough and noisy breathing since 5 weeks of age. He had been evaluated many times in a local emergency department for episodes of cough, wheeze and retractions. His parents reported that his symptoms seemed to be worse when he was supine and did not respond well to steroids or bronchodilators. He had been feeding and growing normally.

On examination he was slightly tachypnoeic with mild subcostal retractions and coarse upper airway sounds. He was admitted for observation and underwent CT scanning of the chest and neck with intravenous contrast followed by three-dimensional reconstruction and virtual bronchoscopy. This revealed a right sided tracheal bronchus arising 1.2 cm above the carina, comparable in size to the right mainstem bronchus (figs 1 and 2).

Scanning was performed on a 64-slice multidetector CT scanner (Siemens Sensation 64) using low dose paediatric protocols for minimum patient dose. Specific scan parameters included CARE Dose software (Siemens) with kV 120 and mAs 60. Slice thickness was 0.75 mm from 0.6 mm detectors and scan reconstruction was at 0.5 mm intervals. The effective radiation dose to a patient of this size can be as high as 4.6 mSv using these parameters. The risks and costs of such a study must be weighed against those of bronchoscopy under sedation, the usual means of making the diagnosis.

An incidence of up to 2% has been reported for tracheal bronchus.[1] The term "pig bronchus" or "bronchus suis" is used when the entire right upper lobe bronchus stems from the trachea. Congenital tracheobronchial abnormalities may be asymptomatic and discovered as incidental findings or may present with stridor, recurrent pneumonia or atelectasis.[2] Expectant management is preferred and surgical resection is typically reserved for patients with severe or persistent symptoms.

The patient was discharged from hospital with medical treatment for gastro-oesophageal reflux. His symptoms resolved over the course of 2 months.

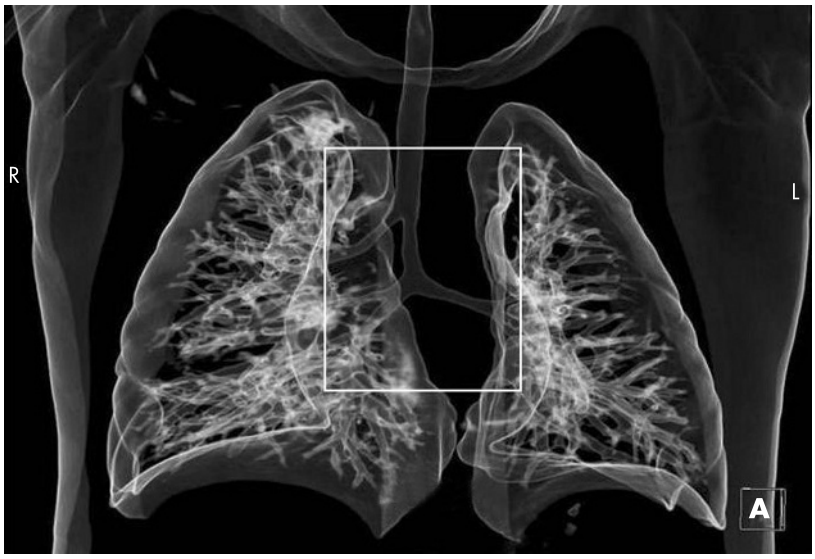


**Figure 1** Three-dimensional reconstruction of airway and lungs from neck and thoracic CT scan showing right sided tracheal bronchus arising 1.2 cm above the carina.

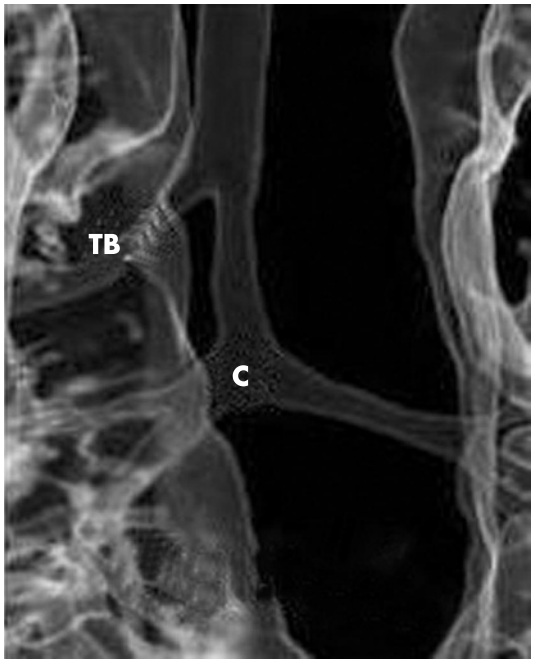


**Figure 2** Detail from box in fig 1 showing a closer view of the tracheal bronchus (TB) and carina (C).

## Learning points

- The differential diagnosis for noisy breathing in infants should include congenital tracheobronchial anomalies, especially when symptoms fail to respond to conventional treatment.
- The ability to diagnose and characterise these abnormalities non-invasively has been enhanced by use of multidetector CT scanning and 3D airway reconstructions.
- Observation of such patients with mild symptoms is an appropriate management option as their symptoms will often improve or resolve with time.

**W A Gower,[1] S A McGrath-Morrow,[1] K D MacDonald,[1] E K Fishman[2]**

[1] Eudowood Division of Pediatric Respiratory Sciences, Johns Hopkins Medical Institutions, Baltimore, Maryland, USA; [2] Russell H Morgan

Department of Radiology, Johns Hopkins Medical Institutions, Baltimore, Maryland, USA

**Correspondence to:** Dr Sharon McGrath-Morrow, Eudowood Division of Pediatric Respiratory Sciences, Johns Hopkins Hospital, 200 North Wolfe Street, Third Floor, Baltimore, Maryland 21287, USA; smorrow@jhmi.edu

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## REFERENCES

1. **Ghaye B,** Szapiro D, Fanchamps JM, *et al*. Congenital bronchial abnormalities revisited. *Radiographics* 2001;**21**:105–19.
2. **Sanchez I,** Navarro H, Mendez M, *et al*. Clinical characteristics of children with tracheobronchial anomalies. *Pediatr Pulmonol* 2003;**35**:288–91.

# Pulmonary puzzle

## ANSWER

*From question on page 41*

Direct inspection of the lesion during video assisted thoracoscopic surgery was undertaken to ensure neither pleural seeding nor pulmonary or diaphragm invasion (see supplementary video). Following this procedure, wide excision of the chest wall tumour was then performed with thoracic cage reconstruction using a titanium mesh. Grossly, the melanoma-like tumour had invaded into the sixth to ninth ribs, adjacent intercostal muscles and overlying chest wall muscles (fig 1A). Microscopically, the tumour consisted of nests and short fascicles of fusiform or polygonal cells with a clear to granular eosinophilic cytoplasm, expanding the parietal pleura with abundant intracellular and extracellular melanin pigmentation (fig 1B). Focal amelanotic tumour cell nests were also present. Immunohistochemically, the tumour cells were reactive to HMB-45 and S-100 protein. The diagnosis of chest wall clear cell sarcoma, formally known as malignant melanoma of soft parts, was then confirmed. Her postoperative course was uneventful. Four courses of adjuvant chemoradiotherapy were administered to this patient. She was disease free at her 10 month follow-up.

Clear cell sarcoma is a rare malignant soft tissue neoplasm histologically composed of nests or fascicles of cells featuring abundant pale staining or clear cytoplasm.[1–3] As a consequence of the tumour's close histological kinship with malignant melanoma, Chung and Enzinger proposed the name "malignant melanoma of soft parts" for this tumour type in 1983.[1] Such a neoplasm occurs mainly among young adults aged 20–40 years as a painless mass or swelling located in the extremities associated with tendons or aponeuroses.[1–3] Nevertheless, tumours do occur in the trunk on rare occasions.[2 4 5] Differential diagnoses include synovial sarcoma, fibrosarcoma, epithelioid form of malignant peripheral nerve sheath tumour, spindle cell melanoma and renal cell carcinoma. One of the main differential diagnoses for clear cell sarcoma is metastatic malignant melanoma, but this was excluded in our patient because no mucocutaneous lesion suggestive of this diagnosis could be found.

Clear cell sarcoma could present as a large chest wall tumour in asymptomatic young adults and should be listed in the differential diagnoses. Early diagnosis and wide excision are essential for a favourable outcome of this tumour.[3]

► A supplementary video is available online only at http://thorax.bmj.com/content/vol63/issue1

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## REFERENCES

1. **Chung EB,** Enzinger FM. Malignant melanoma of soft parts. A reassessment of clear cell sarcoma. *Am J Surg Pathol* 1983;**7**:405–13.
2. **Parasuraman S,** Rao BN, Bodner S, *et al*. Clear cell sarcoma of soft tissues in children and young adults: the St. Jude Children's Research Hospital experience. *Pediatr Hematol Oncol* 1999;**16**:539–44.
3. **Kawai A,** Hosono A, Nakayama R, *et al*. Clear cell sarcoma of tendons and aponeuroses: a study of 75 patients. *Cancer* 2007;**109**:109–16.
4. **Suehara Y,** Yazawa Y, Hitachi K, *et al*. Clear cell sarcoma arising from the chest wall: a case report. *J Orthop Sci* 2004;**9**:171–4.
5. **Hersekli MA,** Ozkoc G, Bircan S, *et al*. Primary clear cell sarcoma of rib. *Skeletal Radiol* 2005;**34**:167–70.

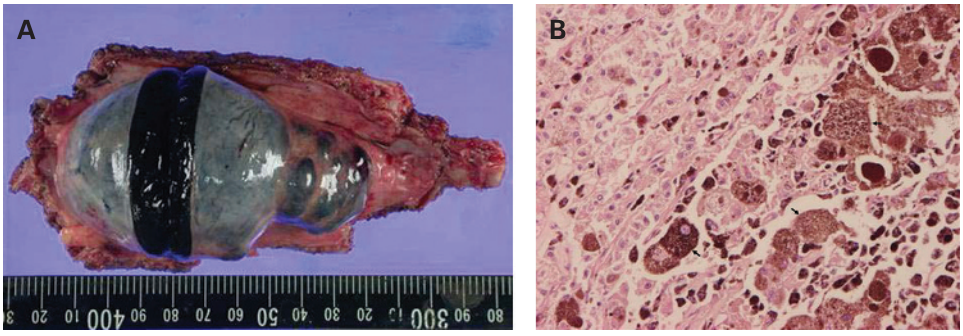


**Figure 1** (A) Grossly, the melanoma-like tumour appeared to arise from the soft tissue of the chest wall with regional rib involvement. (B) Microscopically, focal clear cells and a cellular nesting appearance are noted with abundant melanin pigmentation (black arrow, haematoxylin-eosin, 100× magnification).