

primary osseous dysplasia or pressure-induced responses to neurofibromas^(2,8).

Although the diagnosis of NF1 often relies on cardinal clinical findings, cross-sectional imaging studies can provide valuable information in sundry settings. Particularly for NF1 patients with skull defects, CT is essential for detecting and following up the lesions, given that progressive bone erosion occurs in more than half of all cases⁽²⁾ and such erosion can require calvarial reconstruction with bone grafts or titanium mesh⁽¹⁾. However, progressive bone resorption can predispose to long-term implant instability, the best approach to NF1 calvarial defects therefore remaining undetermined⁽²⁾.

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<http://dx.doi.org/10.1590/0100-3984.2016.0114>

Boerhaave's syndrome: a differential diagnosis of chest and abdominal pain

Dear Editor,

A 61-year-old male patient presented with a two-day history of diarrhea and vomiting, reporting dyspnea, as well as severe lower abdominal and thoracic pain with irradiation to the precordium and left shoulder. Physical examination showed a rigid abdomen and reduced breath sounds, with coarse crackles in both lung bases. Computed tomography (CT) of the thorax showed posterior pneumomediastinum and periesophageal densification

by heterogeneous content (Figures 1A and 1B), and CT of the abdomen, complemented with a small amount of iodinated oral contrast, revealed extravasation to the posterior mediastinum (Figures 1C and 1D). In the emergency room, the patient had another episode of vomiting, which was followed by desaturation. He was immediately transferred to the operating room, where he underwent an extensive surgical procedure, during which esophageal perforation was identified in the distal third of the intrathoracic region. The perforation was closed, and the gastric contents were drained. The patient evolved to hemodynamic instability and was transferred to the intensive care unit.

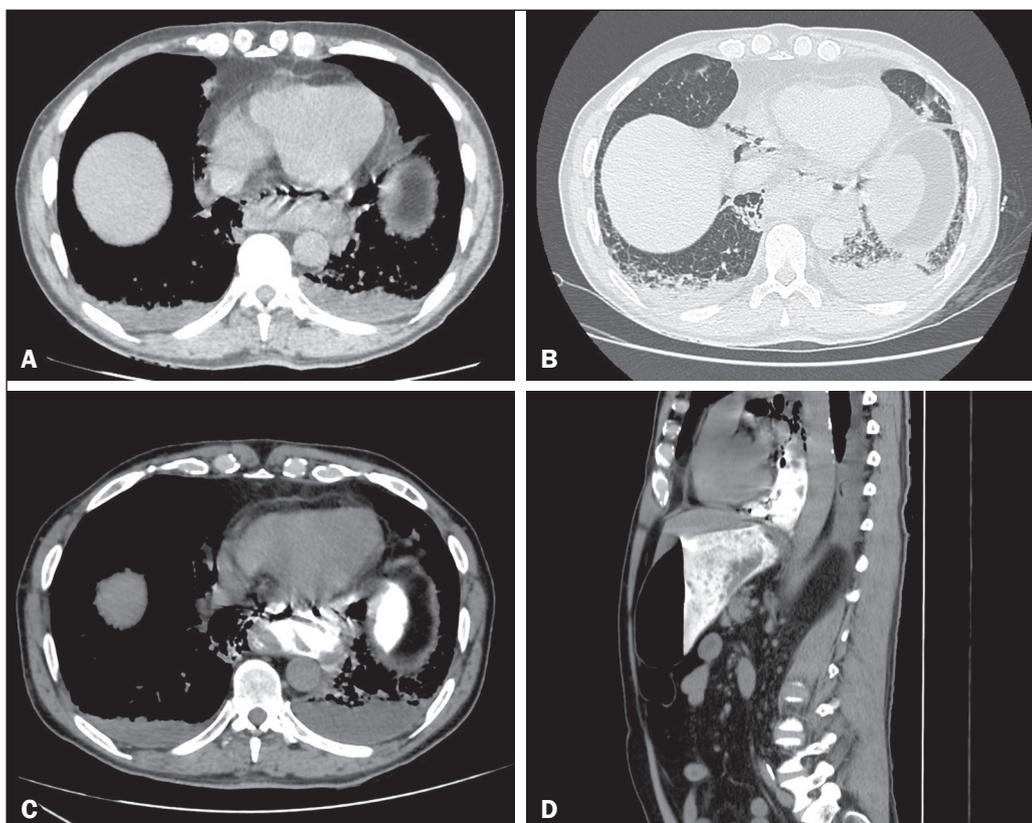


Figure 1. Axial CT of the thorax, with mediastinal and lung window settings (A and B, respectively), showing pneumomediastinum and a collection in the region of the distal thoracic esophagus. Oral contrast-enhanced CT scan of the abdomen, in the axial and sagittal planes (C and D, respectively), showing leakage of ingested material into the periesophageal collection.

Boerhaave's syndrome is the spontaneous perforation of the esophagus resulting from a sudden increase in intraesophageal pressure combined with negative intrathoracic pressure⁽¹⁾. It is a rare condition, with an annual incidence of only 3.1 cases/1,000,000 population. Approximately 15% of esophageal perforations occur spontaneously, and the mortality rate exceeds 40%^(2,3).

Although Boerhaave's syndrome-related perforation occurs most commonly in the posterolateral intrathoracic aspect of the esophagus⁽¹⁾, it can also occur in the cervical and intra-abdominal regions. The condition results in mediastinal contamination by gastric contents, precipitating chemical mediastinitis, with the possibility of evolution to bacterial infection and necrosis⁽⁴⁾.

Patients with Boerhaave's syndrome typically develop signs and symptoms of severe chest pain and subcutaneous emphysema. However, one third of such patients develop atypical symptoms or are admitted to the hospital with respiratory failure or shock^(4,5). Patients with cervical perforations can present with local pain, dysphagia, and dysphonia, as well as tension on sternocleidomastoid muscle palpation and crackles due to subcutaneous emphysema. In addition to Boerhaave's syndrome, the differential diagnoses of chest and abdominal pain should include myocardial infarction, pulmonary embolism, aortic dissection, and pancreatitis⁽¹⁾.

Conventional radiography, barium swallow, and, especially, contrast-enhanced CT are of great value for the timely detection of Boerhaave's syndrome. CT shows the lungs, mediastinum, pleura, and aorta in greater detail, as well as having greater sensitivity in the detection of fluid collections. The findings corroborating rupture include esophageal edema with parietal thickening; perilesional fluid collections with or without a gaseous component; mediastinal widening; and fluid or air in the pleural and retroperitoneal spaces.

In cases of esophageal rupture, the basic therapeutic options include conservative treatment, endoscopic procedures,

and surgery^(6,7). The conservative treatment consists in the interruption of oral food intake, together with fluid administration, enteral nutrition, antibiotic therapy, the use of beta-blockers, and drainage of the perilesional collections. Endoscopic therapy with stent placement can be reserved for cases in which there is early diagnosis, without contamination. Finally, the indication for surgical treatment, which varies from local debridement to the extensive resection of the esophagus, depends on factors such as the extent of the rupture, concomitant diseases, and the presence of contamination or signs of sepsis.

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<http://dx.doi.org/10.1590/0100-3984.2016.0138>

Prenatal diagnosis of an acardiac twin

Dear Editor,

A 32-year-old female patient who was pregnant with twins presented for a regular prenatal checkup with her obstetrician at 25 weeks of gestation. It was her second pregnancy, and she had

carried the first pregnancy to delivery. She was asymptomatic. Ultrasound showed that one twin was morphologically normal and that the other was hydropic, with involution of the brain and only the most rudimentary cardiac tissue (Figure 1).

Recent studies have highlighted the importance of imaging examinations in fetal medicine⁽¹⁻³⁾. Multiple pregnancies are

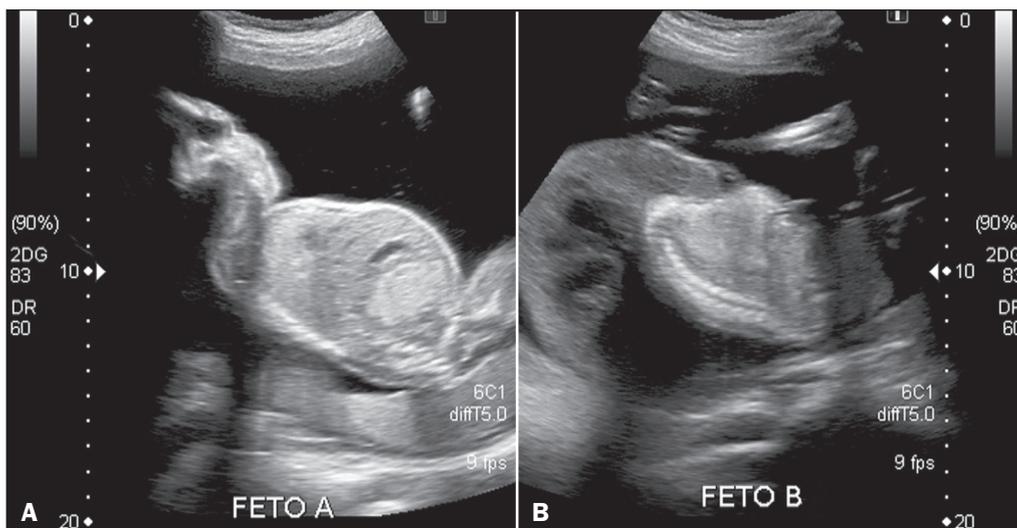


Figure 1. Ultrasound showing comparative images, in the sagittal plane, of a morphologically normal fetus (A) and of a fetus with a bizarre anatomical configuration (B), including the absence of brain formation, no upper or lower limb buds, and hydrops fetalis.