- 14 **Patterson MC**, Pentchev PG. Diagnosis of Niemann-Pick disease type C. *J Pediatr* 1994;**124**:655–6.
- 15 **Yan-Go FL**, Yanagihara T, Pierre RV, et al. A progressive neurologic disorder with supranuclear vertical gaze paresis and distinctive bone marrow cells. *Mayo Clin Proc* 1984;**59**:404–10.
- 16 **Yerushalmi B**, Sokol RJ, Narkewicz MR, et al. Niemann-pick disease type C in neonatal cholestasis at a North American Center. *J Pediatr Gastroenterol Nutr* 2002;**35**:44–50.
- 17 **Kahn E**, Daum F, Markowitz J, et al. Nonsyndromic paucity of interlobular bile ducts: light and electron microscopic evaluation of sequential liver biopsies in early childhood. *Hepatology* 1986;**6**:890–901.
- 18 **Kocak N**, Gurakan F, Yuce A, et al. Nonsyndromic paucity of interlobular bile ducts: clinical and laboratory findings of 10 cases. *J Pediatr Gastroenterol Nutr* 1997;**24**:44–8.
- 19 **De Tommaso AM**, Kawasaki AS, Hessel G. Paucity of intrahepatic bile ducts in infancy—experience of a tertiary center. *Arq Gastroenterol* 2004;**41**:190–2.
- 20 **Yehezkel-Schildkraut V**, Munichor M, Mandel H, et al. Nonsyndromic paucity of interlobular bile ducts: report of 10 patients. *J Pediatr Gastroenterol Nutr* 2003;**37**:546–9.
- 21 **Alvelius G**, Hjalmarson O, Griffiths WJ, et al. Identification of unusual 7-oxygenated bile acid sulfates in a patient with Niemann-Pick disease, type C. *J Lipid Res* 2001;**42**:1571–7.
- 22 **Vanier MT**, Millat G. Niemann-Pick disease type C. *Clin Genet* 2003;**64**:269–81.
- 23 **Guo Y**, He W, Boer AM, et al. Elevated plasma chitotriosidase activity in various lysosomal storage disorders. *J Inherit Metab Dis* 1995;**18**:717–22.

IMAGES IN PAEDIATRICS.....

doi: 10.1136/adc.2006.096941

Extreme subcutaneous emphysema of eyelids

We describe a previously healthy 14 month old girl with a one day history of bilateral periorbital swelling. During the previous evening, parents had noticed slight swelling over the neck and right eye, which during the night worsened and spread to the left eye and chest. There was no history of fever, cough, breathing difficulty, trauma, or surgery; however, she had been constipated for the last four days. On examination she was in respiratory distress with gross swelling of the face and eyelids (fig 1). Her temperature was 100°F, pulse 130, and respiratory rate 65 per minute. Crackles were palpable over the neck and chest; x ray examination showed pneumomediastinum, subcutaneous emphysema of chest, neck, and orbits (fig 2). She was treated conservatively; the swelling subsided considerably during the next 10 days and disappeared completely after six weeks (fig 3).

Orbital emphysema is an uncommon complication of facial injuries or operative procedures.^{1–2} It can also be caused by straining such as the Valsalva manoeuvre or even blowing the nose.³ Since there was a history of constipation in our patient, it was assumed that straining led to interstitial and subsequently orbital emphysema. Emphysema usually resolves spontaneously in 2–3 weeks, therefore no active treatment is needed, even when it is severe.⁴

Figure 1 The patient had extreme swelling of the eyelids; the eyes were shut tightly, and the face swollen.

Figure 2 Skull x ray showing subcutaneous emphysema of the orbits.

M Azam, M Jamal, N Bhatti
Children's Hospital, Pakistan Institute of
Medical sciences, Islamabad, Pakistan;
Morrison85@hotmail.com

Competing interests: none

Parental consent was obtained for publication of the figures

References

- 1 **Ortak T**, Mithat Akan I, Sensoz O. Subcutaneous emphysema of the eyelid and orbital cone after maxillary fracture. *Ann Plast Surg* 2001;**46**:195.
- 2 **Hasel R**, Arora SK, Hickey DR. Intraoperative complications of laparoscopic cholecystectomy. *Can J Anaesth* 1993;**40**:459–64.
- 3 **Mohan B**, Singh KP. Bilateral subcutaneous emphysema of the orbits following nose blowing. *J Laryngol Otol* 2001;**115**:319–20.
- 4 **Zachariades N**, Mezitis M. Emphysema and similar situations in and around the maxillo-facial region. *Rev Stomatol Chir Maxillofas* 1988;**89**:375–9.

Figure 3 Normal face of the child without swelling of eyelids and face.