Delayed-onset right-sided congenital diaphragmatic hernia and group B streptococcal septicemia
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This male infant was delivered vaginally at 37 0/7 weeks of gestation. The course of the pregnancy had been unremarkable except for prolonged rupture of membranes 25 hours prior to delivery. At the age of 6 hours, he developed respiratory distress with tachypnea and grunting. Blood cultures were drawn and antibiotic therapy with amoxicillin and tobramycin was started. Respiratory support with nasal CPAP was necessary. The chest X-ray showed increased non-specific pulmonary markings in the left upper lobe which were interpreted as pneumonic infiltration (Fig. 1). Blood cultures were positive for group B streptococci and antibiotic therapy was continued with penicillin G.

Although clinical and laboratory signs of sepsis resolved completely within the first 3 days of therapy, the patient could not be weaned from nasal CPAP. On day 4 of life, a repeat chest X-ray demonstrated opacification of the right-sided middle and lower lung fields with loss of silhouette of the right hemidiaphragm (Fig. 2). Since ultrasound examination demonstrated no effusion, extensive pneumonia of the right middle and lower lobes was assumed.

On day 10 of life, the patient’s respiratory compromise was unchanged. The chest X-ray now showed an opacification of almost the entire right lung field. Bowel gas was seen abnormally high on the right side suggesting displacement of the liver into the right hemithorax (Fig. 3). Ultrasound examination confirmed the diagnosis
of a right-sided diaphragmatic hernia. The infant was transferred to the pediatric surgery unit where the hernia was repaired successfully and further recovery was uneventful.
CXR on admission: nasal CPAP (PEEP 5 cmH₂O, FiO₂ 0.3) with mediastinal shift.
Fig. 2

CXR on 4th day: nasal CPAP (PEEP 5 cmH₂O, FiO₂ 0.21).
CXR on 10th day: nasal CPAP
(PEEP 5 cmH$_2$O, FiO$_2$ 0.3)
Surprisingly, there are reports of about 30 cases – mainly published in the seventies and eighties – describing an association of delayed-onset diaphragmatic hernia and group B streptococcal septicemia (1,2).

A mere coincidence fails to explain this phenomenon because of two reasons: first, the association with group B streptococcal septicemia is (with one exception) only described in right-sided hernia; second, the combination of delayed diaphragmatic hernia with bacterial septicemia other than group B streptococci has not been reported. Assuming an incidence of right-sided congenital diaphragmatic hernia of 1:20’000 and an incidence of neonatal group B streptococcal septicaemia of 1:330, the probability of a coincidence would be calculated at 1: 6’000’000.

Nevertheless, there is no satisfactory explanation of the described phenomenon (3). Some authors hypothesize, that congenital diaphragmatic hernia leads to reduced motion of the diaphragm at the time of delivery which again predisposes to the development of pneumonia and septicaemia. Others stress the fact that neonatal group B streptococcal septicemia can lead to hepatomegaly, which may prevent the liver from being displaced into the right hemithorax during the first days of infection until septicemia resolves. The delay in herniation could also be explained by the fact that in all reported cases intrathoracic pressure was increased by positive pressure ventilation or – as in our case – by CPAP.
In all published cases, secondary opacification of the right lung on chest X-ray was misinterpreted and right-sided diaphragmatic hernia was diagnosed with considerable delay. Neonatologists should be aware of the association of group B streptococcal septicemia and delayed manifestation of right-sided congenital diaphragmatic hernia.

