Epiglottitis Diagnosed Within Hours of Birth

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(Editorial Comment: The authors suggest that epiglottitis should be considered as a source of respiratory obstruction even in the newborn.)

Acute epiglottitis refers to an acute infectious inflammation of the epiglottis usually associated with infection of the immediately adjacent supraglottic structures. It is a well-recognized cause of catastrophic upper airway obstruction in children and adults.1 *Haemophilus influenzae* type B is widely accepted as being the most common etiologic agent giving rise to epiglottitis in all age groups.2-4 Since the introduction of conjugated vaccines against *H influenzae* type B, there has been a sharp decline in the incidence of invasive diseases caused by this organism in children, including epiglottitis.5-7 Epiglottitis has been recognized as an important, uncommon treatable cause of potentially fatal upper airway obstruction that can occur at almost any age. Cases before 1 month of age are quite rare. The earliest cases reported in the English literature include a case of a 5-day-old baby boy with *Staphylococcus aureus* epiglottitis and pharyngitis,8 and a case of an 8-day-old baby girl with *Streptococcus pyogenes* supraglottitis.9 We present an interesting case of epiglottitis occurring within hours of birth.

**CASE REPORT**

This is the case of a baby girl (gestation: 40 weeks and 6 days) born to a G5, T4, PO, AO, L4 healthy 32-year-old mother of Lebanese descent, who tested negative for hepatitis B (HBs Ag), syphilis (VDRL), group B streptococci (GBS), and rubella. Pregnancy was uncomplicated. There were no maternal infections or infectious contacts noted during pregnancy or in the immediate postpartum period. The mother’s glucose tolerance test was slightly elevated in the last trimester, but no medical intervention was required. The mother’s only regular medication consisted of a daily multivitamin preparation (Materna). Normal ultrasounds were noted at 18 and 28 weeks. Spontaneous vaginal vertex delivery followed 4.5 hours of labor, during which no medications were given. The second stage lasted 15 minutes. There was spontaneous rupture of the membranes 2.5 hours before delivery, at which time the amniotic fluid was clear. APGAR scores at 1 and 5 minutes were 5 and 7, respectively, and no meconium was noted. The baby, weighing 4.130 g, was noted to be grunting with nasal flaring and intercostal retractions at birth. The neonate required bag and mask ventilation immediately after delivery for approximately 1 minute before spontaneous respiratory effort was established. At 10 minutes of age, the infant was pink on room air, despite continued grunting, nasal flaring, substernal retractions, and copious nasopharyngeal secretions. At 12 hours of age, the baby was noted to be increasingly distressed with saturation of 68% in room air. The infant was placed in a humidified oxygen hood with 45% to 50% oxygen. The oxygen saturation consequently improved to greater than 90%. Blood cultures were drawn, and intravenous antibiotics were started (ampicillin and gentamicin).

At 18 hours of age, the infant was transferred to our neonatal intensive care unit with increasing respiratory distress on 100% oxygen. On admission, the infant had a mild elevation of temperature (37.8°C) and a mild leukocytosis (39,900 white blood cells per mL). Because of her inspiratory stridor, a direct laryngoscopy was performed, showing an edematous, erythematous epiglottis bathed in copious purulent secretions. A tracheal aspirate was obtained. The preliminary Gram’s stain showed gram-positive cocci. The baby was subsequently intubated, and empiric antibiotic treatment for acute epiglottitis was initiated (ampicillin, cefotaxime, and clavulanic). After consultation with the infectious disease service, results of a chest x-ray showed minimal interstitial fluid, which cleared up rapidly on follow-up. Echocardiogram results were normal. Tracheal aspirate culture subsequently grew a mixed flora of *β*-hemolytic *streptococcus*, coagulase-negative *staphylococcus*, and *Escherichia coli*. Results of other cultures, including blood, stool, urine, and vireology cultures, were negative. Lumbar puncture was attempted several times, but no cerebrospinal fluid was obtained.

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224
should always be wide, though in our case the limiting factor was the proximity of the brainstem. We advised postoperative chemotherapy to the patient, which he refused.

The prognosis of osteogenic sarcoma of the craniofacial bones is quite poor. Five-year survival rate has been described as 9% for skull bones, which is far worse than 23% to 33% described for maxilla and mandible. A scan of the reported cases of osteogenic sarcoma of the temporal bone showed that of 19 cases only 2 had survived beyond 1 year. However, our case was unusual in having survived beyond 6 years.

However, despite its rarity, this entity should always be kept in mind for cases of any rapidly growing bony swelling in the postauricular region with facial nerve involvement, so that early treatment can be instituted.

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REFERENCES

The neonate tolerated respiratory support of room air and a continuous positive airway pressure (CPAP) of 5. The baby was followed-up via direct laryngoscopy by the pediatric otolaryngology service. Gradual resolution of the epiglottic edema and erythema was noted. After receiving 6 full days of intravenous antibiotics, an increasing air leak was noted around the endotracheal tube. At this point, the neonate was started on a 24-hour course of intravenous dexamethasone. Following extubation in the operating theater, laryngoscopic examination was noted to be completely normal. At this time, resolution of the leukocytosis was noted (white blood cell count 14,700/mL). The baby remained stable on room air with no objective signs of any respiratory distress. Oral feedings were initiated successfully. On day 11, following completion of 10 full days of antibiotics, she was discharged home. The baby has remained well since discharge.

DISCUSSION

Review of the literature shows that this appears to be the youngest patient reported with acute epiglottitis. Our neonate had a very limited inflammation that was almost entirely within the epiglottis. This case report should serve to highlight epiglottitis as an uncommon cause of acute upper airway obstruction in the newborn. When confronted with a neonate with unexplained respiratory distress, particularly if inspiratory stridor or signs of an upper airway obstruction are present, diagnostic examination should include a direct or flexible laryngoscopy performed by a skilled intubator. In performing this vital examination, one should make a diligent visual search for structural causes of upper airway obstruction (e.g., bilateral vocal cord paralysis, laryngeal cyst, web malformation, epiglottitis, and so forth). If significant edema and erythema of the epiglottis are noted in a septic neonate with respiratory distress, the baby should undergo endotracheal intubation and be started on empiric broad spectrum antibiotics. The addition of a short course of steroids has anecdotally been believed to hasten the resolution of the disease. However, no clinical trials have addressed this issue. Hence, the efficacy of steroids remains unproven. Cultures are ideally taken from the epiglottis at time of initial intubation because these most accurately reflect the causative organisms in acute epiglottitis. The disease may be followed by direct laryngoscopy with the endotracheal tube in place, as well as monitoring for the presence of an increasing air leak around the endotracheal tube (signifying resolution of upper airway edema). Extubation should be accomplished in the controlled environment of the operating theater. We have seen a dramatic decrease in the number of cases of epiglottitis in the general pediatric population since the introduction of the conjugated vaccine. However, epiglottitis in the neonate and young infant, before immunization, will continue to be an important, albeit uncommon, cause of acute upper airway obstruction.

REFERENCES

Laryngotracheal Stenosis in a Case of Pena-Shokier Syndrome

Kathleen R. Billings, MD, Marc M. Kerner, MD, James F. Padbury, MD, and Elliot Abemayor, MD, PhD

(Editors' Comment: The authors alert the community of physicians of the potential association of subglottic stenosis of this rare congenital syndrome.)

The Pena-Shokier Syndrome is a rare, lethal disorder first described in 1974. It is characterized by severe camptodactyly, clubfoot, knee and hip ankylosis, facial anomalies, and pulmonary hypoplasia. A myriad of other anomalies have been described in the 59 cases reported to date, including hypertelorism, low-set malformed ears, depressed nasal tip, small mouths, and talipes equinovarus, to name a few. This report describes a newborn child with Pena-Shokier Syndrome who required emergent airway intervention related to laryngotracheal stenosis and hypoplastic lungs. This is only the second case to describe laryngeal stenosis in a neonate with this disorder.

CASE REPORT

A GI, PI 26-year-old gave birth to a 37 weeks' gestation boy via Cesarean section. The mother was noted to have polyhydramnios and decreased fetal movements on ultrasound. Prenatal ultrasound had revealed the presence of a Dandy-Walker cyst in the posterior fossa and an absent corpus callosum. At delivery, the baby was noted to have an Apgar score of 5 at 1 minute and 7 at 5 minutes. Soon after birth, the baby was in respiratory distress and attempts to intubate the baby over a neonatal 2.5-mm bronchoscope were unsuccessful. At bronchoscopy, the laryngeal introitus and supraglottis appeared to be a mass of tissue. There was a slit-like opening that was presumably the glottis, but it was unable to accommodate the bronchoscope. A tracheotomy was performed and an 8.0 Shiley neonatal tracheostomy tube was inserted. The larynx was markedly abnormal (Fig 1).

After obtaining an airway, the baby was transferred to the neonatal intensive care unit where a chest x-ray showed bilateral pleuropneumothoraces and underdeveloped lungs. A complete work-up of the baby over the next 48 hours showed many features consistent with Pena-Shokier syndrome including syndactyly, pulmonary hypoplasia, clubfoot, low-set ears, and malformed facial features. Because of the neurological defects, prolonged anoxia, and pulmonary hypoplasia, ventilatory support was withdrawn. The baby expired 48 hours after birth, and an autopsy was performed.

Findings at autopsy showed multiple congenital anomalies consistent with the Pena-Shokier phenotype. These included pulmonary hypoplasia with a lung weight of 26 g (normal, 66 g), distal arthrogryposis, malformed facial features (micronathia, ocular hypertelorism, low-set ears), and clubfoot. The trachea was narrowed to 0.1 cm at 2 cm above the carina. A small, narrow glottis was noted (Fig 2). Developmental abnormalities of the brain included Dandy-Walker cerebellar agenesis, corpus callosum agenesis, corticospinal tract dysgenesis, and pontomedullary dysgenesis.

DISCUSSION

In 1974, Pena and Shokier described an unusual syndrome in two female siblings consisting of camptodactyly, clubfoot, knee and hip ankylosis, facial anomalies, and pulmonary hypoplasia. Since this initial report, 57 additional cases have been reported in the literature describing a variety of associated features. Despite this wide array, the most frequent cause of death in these infants is pulmonary hypoplasia. This report describes the unusual added feature of laryngeal stenosis. This finding was noted in one prior case.

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