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Letters to the Editor

Congenital Hydrops and WPW Syndrome

Congenital Wolff-Parkinson-White (WPW) syndrome, intrauterine tachycardia, heart failure, and hydrops fetalis is an unusual sequence of events bringing a premature baby to a neonatal intensive care unit (ICU).

Case Report

The mother, a 24-year-old primigravida, noticed a sudden disappearance of fetal movements at the 34th week of an uneventful pregnancy. The next day, the fetal heart rate was found to be about 200/min and she was admitted. A few hours later the tachycardia rose to 240/min, prompting preparations for an emergency cesarean section, as cardiac origin of the fetal tachyarrhythmia was suspected. At the time of maternal anesthesia the fetal heart rate dropped suddenly to 156/min. There was no hydramnios and the placenta (434 g) was normal. A 2480-g male infant was delivered, Apgar 3–7–8. He was cyanotic with bradycardia and no spontaneous respiration. He was intubated immediately and ventilated with pure oxygen. His color improved rapidly and he showed spontaneous movements after 4 min. He was kept intubated and ventilated during referral to our neonatal ICU.

On admission the baby was hydropic (generalized body edema), heart rate was 134/min, blood pressure 48/20 mmHg, and temperature 36.4°C. There was no heart murmur. Physical examination of the abdomen revealed massive edema and ascites. The liver was 2 cm below the right costal margin and the spleen was not palpable. No obvious malformations were seen. Neurologically the infant appeared normal.

A moderately enlarged heart, signs of pulmonary perivascular edema, fluid in the lung fissures, considerable bilateral pleural effusions, and ascites were found on x-ray examinations.

Arterial blood gases were: pH 7.26, $PaCO_2$ 31 mmHg, BE −13.0 mmol/liter, PaO_2 73 mmHg (FIO_2 1.0); total serum proteins 42 g/liter; all other investigated biochemical parameters were within the normal range. Results of bacteriological and virological examinations remained negative. An ECG showed a typical WPW pattern and echocardiography was normal.

The infant remained intubated and ventilated with FIO_2 over 0.8 during the first 24 h. Edema was treated with salt-poor albumin and furosemide. The weight dropped from 2480 g to 2010 g. Arterial blood gases improved as edema decreased and at 58 h of age the child was extubated. During the first two weeks of life the heart rate remained between 120 and 156/min. At the age of 14 days, the first extrauterine episode of supraventricular tachycardia with a heart rate of 300/min was noticed. Digitalization brought back sinus rhythm 2 h after the loading dose. The further history was uneventful and he was discharged home on day 24 (weight 2470 g).

He was checked by us three and six months later. The WPW pattern still present at three months had disappeared at six months and digoxin was discontinued. At the age three years he had age-corresponding psychomotor development. ECG and echocardiograms were normal.

The prevalence of nonimmune hydrops fetalis (NIHF) has been estimated to be at least 1.4 cases for each 10,000 deliveries [2]. Since the introduction of anti-D prophylaxis, immune hydrops is much rarer. The NIHF may well have become the most common form to be diagnosed and treated in the neonatal ICU [2]. NIHF can result from a large number of fetal, placental, and maternal causes [2]. In 1969, NIHF was associated with intrauterine tachycardia for the first time [9]. This special group deserves attention because of the following characteristics:

1) The diagnosis may not be evident; fetal tachycardia is usually interpreted as the result of fetal distress and not as the cause [7]. Repeated cardiotocographic registrations, the oxytocin challenge test, fetal ECG recording [1, 7], and fetal echocardiography [6] may be helpful in differentiating the origin of fetal tachycardia. It is of outstanding importance to bear in mind the possibility of an underlying fetal cardiac defect in the presence of intrauterine tachyarrhythmia, as the heart rate may become normal during or shortly after delivery ([9] and our case).

2) The etiology of congenital tachyarrhythmias remains often unknown. They have been associated with a variety of congenital heart diseases [3], but most of the cases have no malformation [7] and different possible mechanisms leading to supraventricular tachyarrhythmias have been observed [4]. The WPW syndrome as the underlying cause has been recognized in a few instances [7].

3) Congenital hydrops due to paroxysmal intrauterine tachycardia can be successfully managed by conventional neonatal intensive care techniques. Survival rates in newborns with NIHF and without congenital cardiac malformations should be high even when hydrops is severe.

4) The outcome is better if the diagnosis of heart rate disturbance is made antenatally [8] and when

the delivery can be put forward and undertaken in collaboration with a neonatal intensive team [2]. In a few cases, prenatal treatment of fetal paroxysmal tachyarrhythmia has been successful [5].

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Asplenia Syndrome Complicated by Purulent Pericarditis

Asplenia syndrome is a disease characterized mainly by congenital absence of the spleen and severe complex heart disease [2]. It is recognized that patients with this syndrome have increased susceptibility to severe infections such as septicemia [3], but there have been no reports of proven purulent pericarditis. We present here a case of asplenia syndrome complicated by purulent pericarditis, which followed wheezy bronchitis and septicemia due to *Staphylococcus aureus*.

A seven-month-old girl was admitted to hospital on 17 September 1983 with high fever and progressive dyspnea of one day's duration.

She had been found to have asplenia syndrome at two weeks of age. The heart, examined by two-dimensional echocardiography, consisted of a single ventricle, a common atrioventricular canal, atrial septal defect, persistent ductus arteriosus, transposition of the great arteries, and probable pulmonary stenosis. Despite medication with digoxin her weight gain was poor, and cardiomegaly progressed gradually.

On admission, wheezing and prolonged expirations were observed. Physical examination revealed: body weight, 5.28 kg; temperature, 39.1°C; heart rate, 182/min; respiratory rate, 62/min. The chest x-ray showed moderate increase of pulmonary markings. The cardiothoracic ratio was 64. Laboratory data were: red cell count, 525 million/mm^3; hemoglobin, 16.0 g/dl; white cell count, 21,800/mm^3 with 87% neutrophils; CRP, 5(+); electrolytes, normal; liver and renal function tests, normal. She was treated for wheezy bronchitis. Fever, tachycardia, and dyspnea were improved by the fourth day of admission, but recurred three days later, and then cardiomegaly progressed. On the 12th day of admission, the blood culture was positive for *Staphylococcus aureus*. On the 17th day of admission, the chest x-ray revealed an almost completely radiopaque left hemithorax (Fig. 1). Computerized tomography of the chest demonstrated that the opacification was due not to atelectasis but to pericardial effusion. No friction rub was ever heard. The ECG showed decreased voltage of QRS and T waves. Pericardiocentesis yielded about 40 ml of thin purulent fluid with Gram-positive cocci, but culture was negative. Treatment with large doses of cephalothin (300 mg/kg/day) was started, and clinical findings were markedly improved within three days. She was discharged 40 days after admission.

Although the typical friction rub is the most reliable diagnostic clue, it is usually absent, as in this case, in purulent pericarditis, unlike other types of pericarditis [1]. If purulent pericarditis occurs in patients with severe heart disease and preceding infection, the diagnosis must be very difficult because other clinical features such as fever, tachycardia, tachypnea, and cardiomegaly may be confused with those of congestive heart failure due to serious infection [1].

Another radiological finding of pericarditis is the so-called globular-shaped heart shadow. In this case, however, there was a different form of heart shadow, confined to the left hemithorax, almost completely occupying it (Fig. 1). It looked more like massive atelectasis of the left superior lobe, superimposed on the heart shadow, rather than pericarditis.

If purulent pericarditis is not treated, the mortality rate is 100% [1]. To avoid missing purulent pericarditis it should be suspected whenever cardiomegaly is observed in a patient with underlying heart disease and serious respiratory infection, even though the cardiac silhouette has not the typical globular shape.