Infant with rapidly progressive respiratory distress

Stefano Pintaldi,[1] Alessia Giuseppina Servidio,[1] Marco Bobbo,[2] Daniela Chicco,[2] Lorenzo Calligaris,[2] Egidio Barbi,[1,2] Giorgio Cozzi [2]

A 7-month-old girl presented to the emergency department with a 12-hour history of difficult breathing. The girl was alert but with a severe tachy-dyspnoea, chest retractions and nodding. Vital signs showed: respiratory rate 70/min, pulse rate 150/min and oxygen saturation 92% on room air. At chest auscultation, mild bilateral basal crackles were noted. Repeated albuterol inhalations, systemic steroid and oxygen administration through high flow nasal cannula were administered, but respiratory distress worsened in the next 2 hours. Capillary blood gas analysis showed: pH 7.37 pCO_2 27 mm Hg, HCO_3 17 mm Hg. A chest radiograph was performed (figure 1).

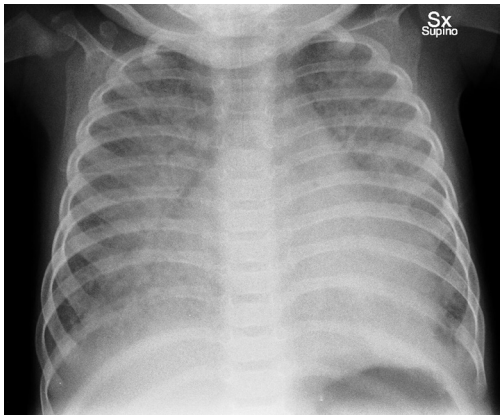

Figure 1 Chest X-ray shows enlargement of the cardiac silhouette with abnormal lung fields characterised by alveolar oedema, air bronchogram on the right side and left basal pleural effusion.

QUESTIONS

1. Which is the most likely diagnosis in this patient?
   - A. Bronchiolitis
   - B. Acute respiratory distress syndrome
   - C. Acute heart failure secondary to dilated cardiomyopathy
   - D. Diabetic ketoacidosis
2. Which test may help to confirm the diagnosis?
3. How should this patient be treated?

ANSWERS TO THE QUESTIONS ON PAGE 369

Answer for question 1

Acute heart failure (AHF) with pulmonary oedema in an infant with dilated cardiomyopathy (DCM). AHF is rare in children (incidence 0.87–7.7/100 000)[1] and should be suspected in case of severe respiratory distress without a clearly pathological pulmonary auscultation, no response to treatment for viral respiratory infections and cardiomegaly at the X-ray. DCM is a leading cause of AHF in children with a peak incidence in the first year of life, and it was diagnosed through echocardiography also in this case.[2 3] It can be either a primary or acquired condition, and the most frequent aetiologies are infectious myocarditis, neuromuscular disorders, inborn errors of metabolism and toxic-related forms. The clinical presentation of DCM may be abrupt, characterised by respiratory distress and poor feeding, similarly to common respiratory infections. Another cause of AHF may be the late presentation of a large L-R shunt sometimes in association with a viral illness (ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage), in which chest X-ray would show pulmonary plethora in addition to oedema. In infants, clinical signs of AHF, as hepatomegaly, jugular vein distention and pulmonary rales can be absent or hard to recognise, so the suspicion should be high.[4]

Answer for question 2

Chest X-ray findings almost invariably include cardiomegaly and may demonstrate pulmonary oedema, infiltrates or pleural effusions making it a fast, reliable and affordable investigation[5] (figure 2). Electrocardiography is a vital diagnostic tool and contributed to the diagnosis also in our case (figure 3). It may reveal signs of ventricular overload or ischaemia (ST-T wave changes) in case of myocarditis or a tachycardia as the cause of cardiomyopathy (ie, atrial ectopic tachycardia or permanent junctional reciprocating tachycardia). Eventually, echocardiography, which can show systolic/diastolic dysfunction and reduction of ventricular ejection fraction, is warranted to confirm the diagnosis. In adjunction, blood tests may show metabolic acidosis and elevation of brain natriuretic peptide.

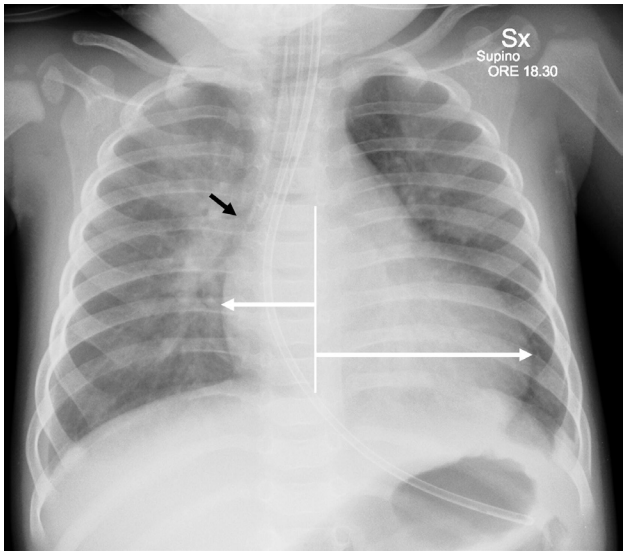

Figure 2 Chest X-ray, after stabilisation, shows partial resolution of alveolar oedema which revealed clearly the cardiac enlargement (white arrows). Endotracheal tube, which was distally displaced (black arrow), was promptly repositioned.

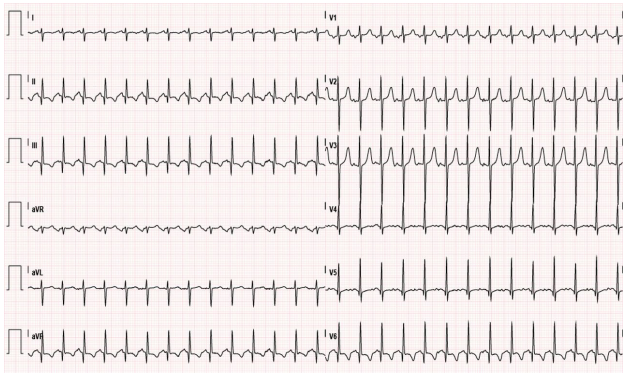

Figure 3 The ECG showed sinus tachycardia (167 bpm), Q waves in inferolateral leads and pathological features of biventricular overload: positive T waves in right precordial leads, negative T waves in inferolateral leads. These signs confirmed the suspect of an underlying cardiac disease.

Answer for question 3

In the setting of AHF, the stabilisation of critical patient is of primary importance. Treatment may include pharmacological options such as diuretics, inotropic agents, anticoagulation and antiarrhythmic. Diuretics should be given as soon as possible in acute symptomatic cardiac decompensation to decrease fluid overload and cardiac work, in adjunction to fluid restriction. Inotropes, including phosphodiesterase inhibitors (milrinone) or adrenergic agonists (dobutamine or dopamine), should be administered when signs of left ventricular dysfunction with poor organ perfusion are present. The use of anticoagulation is usually indicated if the ejection fraction is severely decreased or in the setting of atrial arrhythmias, arterial or venous thromboembolism. Patients with evidence of hypoxaemia and respiratory acidosis may require ventilatory support with positive pressure and mechanical ventilation. In most severe cases that show signs of poor perfusion despite optimal medical treatment, mechanical circulatory support with left ventricular assist device or extracorporeal membrane oxygenation may be necessary as a bridge to cardiac transplantation.[6–9]

Stefano Pintaldi,[1] Alessia Giuseppina Servidio,[1] Marco Bobbo,[2] Daniela Chicco,[2] Lorenzo Calligaris,[2] Egidio Barbi,[1,2] Giorgio Cozzi [2]
[1]University of Trieste, Trieste, Friuli-Venezia Giulia, Italy
[2]Institute for Maternal and Child Health-IRCCS "Burlo Garofolo", Trieste, Italy

Correspondence to Dr Stefano Pintaldi, University of Trieste, Trieste 34127, Italy; pintaldistefano@gmail.com

Contributors SP, AGS and GC wrote the paper. LC, MB and DC supervised the work. EB supervised the work and approved the final revision.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

To cite Pintaldi S, Servidio AG, Bobbo M, *et al*. *Arch Dis Child Educ Pract Ed* 2022;**107**:369–371.

REFERENCES

1 Shaddy RE, George AT, Jaecklin T, *et al*. Systematic literature review on the incidence and prevalence of heart failure in children and adolescents. *Pediatr Cardiol* 2018;39:415–36.
2 Burch M, Runciman M. Dilated cardiomyopathy. *Arch Dis Child* 1996;74:479–81.
3 Nugent AW, Daubeney PEF, Chondros P, *et al*. The epidemiology of childhood cardiomyopathy in Australia. *N Engl J Med* 2003;348:1639–46.
4 Lipshultz SE, Sleeper LA, Towbin JA, *et al*. The incidence of pediatric cardiomyopathy in two regions of the United States. *N Engl J Med* 2003;348:1647–55.
5 Greenwood RD, Nadas AS, Fyler DC. The clinical course of primary myocardial disease in infants and children. *Am Heart J* 1976;92:549–60.
6 Hunt SA, Abraham WT, Chin MH, *et al*. 2009 focused update incorporated into the ACC/AHA 2005 guidelines for the diagnosis and management of heart failure in adults a report of the American College of cardiology Foundation/American heart association Task force on practice guidelines developed in collaboration with the International Society for heart and lung transplantation. *J Am Coll Cardiol* 2009;53:e1–90.
7 Canter CE, Simpson KE, Simpson KP. Diagnosis and treatment of myocarditis in children in the current era. *Circulation* 2014;129:115–28. [Published correction appears in Circulation. 2016 Jan 19;133(3):e30. Simpson. Kathleen P [corrected to Simpson, Kathleen E]].
8 Price JF. Congestive heart failure in children. *Pediatr Rev* 2019;40:60–70.
9 Lasa JJ, Gaies M, Bush L, *et al*. Epidemiology and outcomes of acute decompensated heart failure in children. *Circ Heart Fail* 2020;13:e006101.