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Case Report

Primary Chylopericardium in Preterm Newborn Presenting with Cardiac Tamponade and Obstructive Shock: A Case Report

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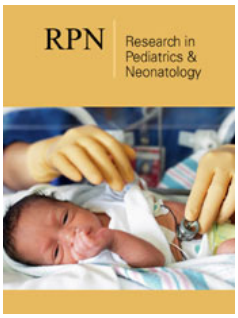

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Abstract

Idiopathic chylopericardium in neonates is exceedingly rare, often benign and presents with varied symptoms. We present a case of obstructive cardiogenic shock due to chylous pericardial effusion in a newborn with fetal hydrops without congenital heart disease. The premature newborn exhibited signs of systemic congestion and respiratory distress few days after birth. Despite initial stability, the infant later deteriorated into cardiogenic shock, prompting emergency echocardiography revealing cardiac tamponade. Emergency pericardiocentesis resulted in immediate hemodynamic improvement. Subsequent management included octreotide therapy and low-fat parenteral nutrition. Laboratory findings confirmed chylous effusion. Conservative management led to resolution without recurrence. This case highlights the importance of early recognition and prompt treatment in neonatal obstructive shock caused by cardiac tamponade. Regular follow-up is crucial to monitor for long-term complications.

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Keywords: Chylopericardium; Cardiac tamponade; Obstructive shock

Introduction

Idiopathic chylopericardium at birth is a very rare event, usually of benign cause, which can have variable presentation and has never been described in the newborn as an acute event causing cardiac to cardiac tamponade without prodromes. We present a case of obstructive cardiogenic shock due to cardiac tamponade caused by chylous pericardial effusion in a newborn with fetal hydrops without cardiac congenital pathology associated.

Clinical case

A premature newborn (29 weeks gestation, weight 2,002kg, length 41cm) is admitted in the Neonatal Intensive Care Unit (NICU) of our hospital after urgent caesarean section performed due to suspected fetal hydrops and cervical shortening. At birth, the patient had an Apgar score of 5 at 1 minute, 7 at 5 minutes and 8 at 10 minutes, with severe difficulty in maintaining

autonomous respiratory drive despite non-invasive ventilatory support, prompting transfer to the NICU. Her presentation upon admission showed signs of systemic congestion with swelling of the lower limbs, severe edema of the trunk, abdomen, and neck, with pitting edema in the explored areas. Chest objective assessment revealed moist pulmonary sounds suggestive of pulmonary overload. Vital signs showed a heart rate compatible with age, normal body temperature, and respiratory rate within normal limits. Venous access was established via umbilical venous catheter (UVC). Emogas analysis performed from UVC at approximately 30 minutes of life showed values of pH, PCO2, and pO2 within normal limits with a maximum support of FiO2 50%. Chest X-ray showed proper positioning of the UVC without gross alterations of lung parenchyma and pleural spaces, together with a normal cardiac silhouette. The electrocardiogram (EKG) showed sinus tachycardia within normal limits for age, PR within normal range, and right-sided predominance with signs of right ventricle overload. Due to hypoventilation, endotracheal surfactant administration was attempted using the LISA technique.

On the initial echocardiogram (Figure 1a) performed two hours after birth, hypertrophy of the interventricular septum (IVS) and the free wall of the right ventricle with normal function were noted. The left ventricle showed normal function, the ductus arteriosus was patent with bidirectional flow in the absence of signs of aortic coarctation. No congenital anomalies were observed, there was no evidence of pericardial effusion.

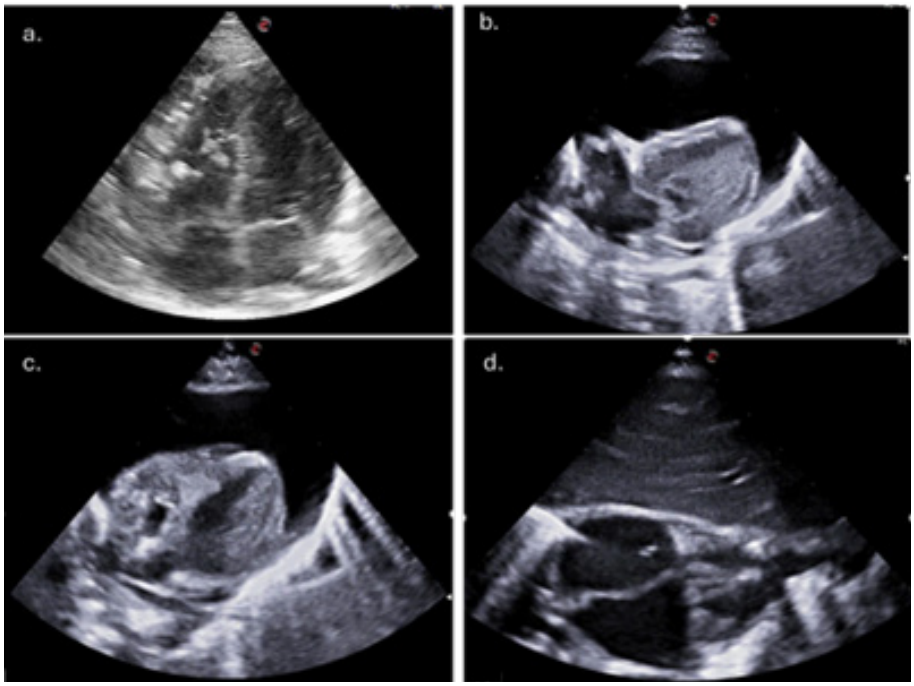


Figure 1: Echocardiography images before, during and after cardiac tamponade. (a) Four chamber view at arrival in newborn intensive care unit; (b) and (c) Images during cardiac tamponade with compression of the right ventricle free wall and "sickle effect" of left atrial roof (partial collapse during the whole cardiac cycle); (d) Subxyfoid view one hour after drainage positioning.

Two days after birth the newborn, following a crying episode, presented tachypnea, cyanosis, and desaturation despite receiving 100% FiO2. The patient was in a state of marked hypoperfusion (blood pressure (BP) 38/13/22mmHg), followed by cardiorespiratory arrest necessitating resuscitative manoeuvres and orotracheal intubation. After fluid resuscitation and sedation with Fentanyl, spontaneous circulation was restored. The heart rate was tachycardic compared to age limits, cardiac tones were muffled. Suspecting cardiogenic shock, an emergency echocardiogram was performed, revealing tamponade pericardial effusion with floating and swinging heart together with severe hemodynamic compromise, compression of the right atrium and of the right ventricle (Figure 1b & 1c). Emergency pericardiocentesis was performed under cardiothoracic surgical assistance and echocardiographic guidance, using anterior percutaneous approach through the apical window with a 22G needle. Eight millilitres of milky fluid were aspirated. The patient showed an immediate improvement in hemodynamic with restoration of adequate peripheral perfusion (BP 55/35/43mmHg), respiratory parameters, and normalization of instrumental findings, with almost complete disappearance of circumferential effusion. The drainage catheter was left in place after X-ray positioning control (Figure 2) with further intermittent removal of 8.5cc over the next 24 hours due to recurrence of intrapericardial fluid. Suspecting chylopericardium, therapy with Octreotide 1mcg/kg/h was initiated, along with low-fat total parenteral nutrition (TPN) with medium-chain triglycerides (MCT) and high protein, antibiotic prophylaxis with Vancomycin was administered.

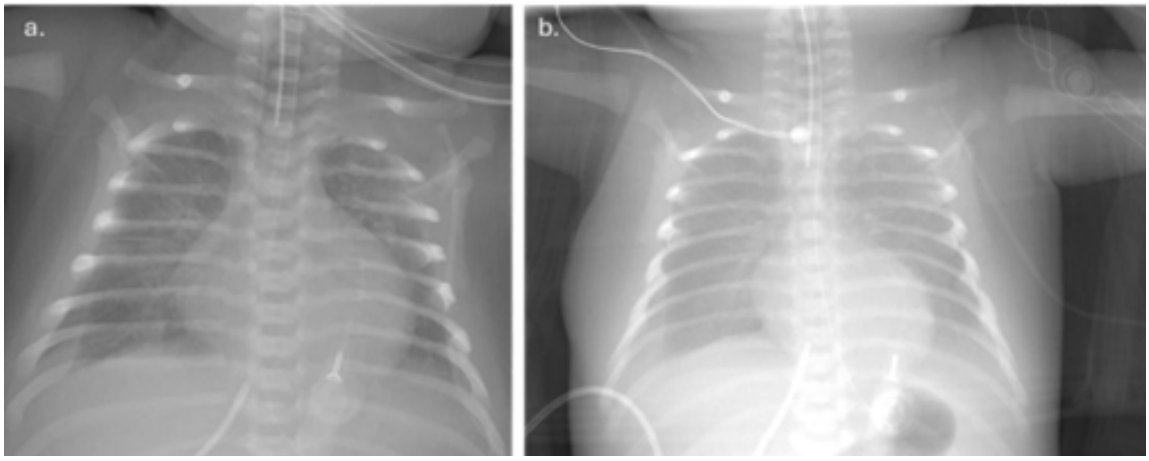


Figure 2: X-Ray images. (a) X-ray recorded just after positioning of pericardial drainage. Cardiomegaly is shown; (b) X- ray recorded after complete drainage of chylous pericardial liquid.

Confirmatory data from the laboratory showed elevated triglyceridamie together with lymphocitosis in the aspirated pericardial fluid with negative cultures. Due to initial recurrence of pericardial effusion, the drainage was set to gravity drainage. In the following days hemodynamic remained stable, the patient had preserved active diuresis and simultaneous improvement in congestive and respiratory symptoms. Additionally, an abdominal ultrasound was performed, revealing ascitic fluid in all exploratory recesses without organ abnormalities while chest ultrasound, later confirmed by chest X-ray, showed bilateral pleural effusion without indication for drainage.

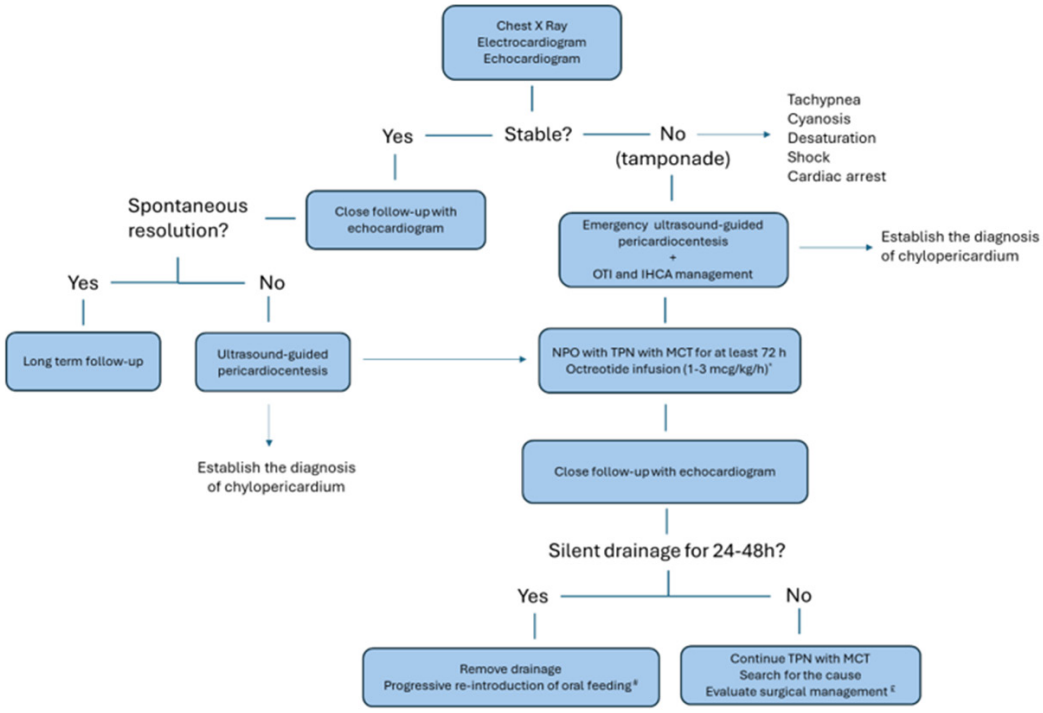
The collection of pericardial fluid in the subsequent days amounted to a total of 11cc. Hematic parameters showed increased cholestasis indices with hormonal balance of various pituitary axes within normal limits. Serology did not show positivity for the main viruses tested (Parvovirus, CMV, EBV). After UVC haemocultures positivity for Staphylococcus Epidermis the patient underwent an antibiotic cycle according to the antibiogram, until cultures turned out negatives. Genetic analyses yielded negative results.

Five days after the event pericardial drainage was closed, as the cardiosurgeon advised, after 2 days of silent drainage and concurrent negativity for effusion when the echocardiogram was performed. Subsequently, under close ecoguided monitoring, drainage removal was performed 72 hours after closure. From a nutritional point of view, after 5-7 days of TPN with MCT, oral nutrition was reintroduced with special milk enriched with MCT, while de-escalating TPN. At the same time, Octeotride has been gradually discontinued. Follow-up echocardiography showed resolution of effusion (Figure 1d), even after reintroduction of complete enteral nutrition, with physiological normalization of right ventricular hypertrophy. Abdominal and thoracic serous cavities also experienced reduction until gradual disappearance of effusions. After extubation and non-invasive support, effective initiation of regular autonomous respiratory mechanics was observed until weaning off oxygen. The patient was followed up by neurologists who did not detect any post-arrest cerebral sequelae following close monitoring. She was guarded with multiple reassessments, transferred to sub-intensive and regular ward care, and lately discharged with ursodeoxycholic acid and vitamin supplementation.

Discussion

The accumulation of chylous fluid in the pericardial cavity of the newborn is a very rare entity, defined as chylopericardium [1-3]. This condition, although rarely described in the neonatal period, can lead to cardiac tamponade up to obstructive shock, representing a life-threatening disorder in newborns. Both in paediatric and adult age, the origin of chylopericardium appears to be secondary to abnormal or damaged lymphatic vessels (e.g., trauma, surgical intervention, viruses, tumours) or increased pressures of the thoracic duct with chylous reflux into the pericardium (e.g., pulmonary hypertension, subclavian vein thrombosis) [4-6]. Rare causes in infants could be linked to lymphatic malformations, as described in the 2018 revision of the International Society for the Study of Vascular Anomalies (ISSVA) Classification [7,8]. The largest sample size retrospective study conducted to date regarding chylothorax in the paediatric age group has shown a high incidence of post-surgical chylothoraxes (91%) with the remaining forms being idiopathic (6%) or congenital (3%), while the exact incidence of chylopericardium in the paediatric population is not known [9]. A recent systematic review on the subject found idiopathic/viral aetiology to be the most frequent in the general population (60%). The diagnosis of this condition follows the observation of cardiac enlargement on chest X-ray and the presence of pericardial effusion on echocardiography with aspiration of milky fluid, finding Triglycerides >500mg/dL and a cholesterol/triglyceride ratio <1 with a predominance of lymphocytic count [6]. In cases of tamponade, the patient may present with muffled heart sounds, paradoxical pulse, Kussmaul's sign with jugular venous distention, tachycardia and tachypnea, as well as pallor and cyanosis. Given the rare nature of the pathology, there are no consensus guidelines reported in the literature regarding the systematic management of chylopericardium in the infant and neonatal population, except in the

case of post-surgical chylothorax in simple and complex congenital pathologies [10]. Furthermore, considering the rare expression of the pathology in the acute stage as seen in our patient, there is no described standard approach, which should rather be linked to clinical judgment. Typically, in the majority of cases of pericardial effusion of unknown origin the initial approach is conservative with close monitoring of patient vital parameters together with serial echocardiography to study the progression of pericardial effusion. In cases of significant pericardial effusion, hemodynamic impact at echocardiography monitoring or tamponade, evacuation is recommended, followed by continuous or intermittent drainage, based on monitoring and clinical assessment [1,3,11]. In case of rare presentation with obstructive shock and circulatory arrest, it is crucial to promptly diagnose and set the treatment within the shortest possible timeframe [12,13]. The simultaneous administration of fluids for volume replacement and evacuation of the obstructive fluid are the two key components for circulatory support and resolution of the primary cause [2]. Drainage pericardial window should always be guided by ultrasound, with puncture in the area where there is the greater amount of fluid [14]. During and after cardiac arrest, comprehensive patient management should not be overlooked, including airway protection and temperature management, close monitoring of vital parameters and serial reassessments. After achieving stable hemodynamic parameters, as complete stabilization is accomplished, in cases of suspected chylous accumulation, parenteral nutrition with low in fatty acids with medium-chain triglycerides (MCT) should be initiated, and therapy with Octreotide should be started in accordance with the latest guidelines [2]. When infections are suspected or confirmed, management through antibiotic therapy is important, while in inflammatory cases the use of steroids should be considered. Our centre has drawn up a protocol to apply in cases of suspected chylopericardium in newborns (Figure 3). Our patient benefited from conservative treatment without subsequent recurrences, even following the restoration of normal nutrition (initially with milk rich in MCT). If a reduction in daily drainage is not achieved, surgical intervention with ligation of the thoracic duct should be considered [6]. If conservative treatment is not conclusive or there are long-term recurrences, in pediatrics population MR guided lymphography is the most useful imaging technique to exclude malformations, even if there are some evidences on liphoscintigraphy [15-17]. TC can be used to exclude mediastinal tumors while more sensitive examinations for the study of obstructions, abnormalities, leaks or fistulas are lymphangiography and lymphscintigraphy with Tc99-sulphur colloid, but the techniques are better described for the adult population [5]. It is not to forget that, given the possible relationship with syndromes such as Noonan or Kippel Trenaunay, as well as the genetic component, it is important to screen patients through genetic analysis [11,17].



* At least for 7 days

#Oral nutrition is to be reintroduced at least after 3 days of TPN with low-fat medium chain tryglyceride (MCT). We gradually start oral feeding with diet rich in MCT together with carbohydrate and protein-based diet. If the patient is stable, after 7 days of Octreotide therapy gradual decalage is to be considered.

£Percardiectomy or pericardio-pleural window or pericardio-peritoneal window ± thoracic duct ligation.

IHCA: intra-hospital cardiac arrest; NPO: Nil Per Os (fasting); OTI: orotacheal intubation; TPN: total parenteral nutrition.

Figure 3: Diagnostic and Therapeutic protocol for newborn with suspected chylopericardium in our centre.

Conclusion

The clinical case described illustrates a very rare event, such as spontaneous neonatal chylopericardium in a patient with suspected fetal hydrops without congenital heart disease, in the absence of causes such as trauma or surgery, with acute onset. In the specific case of tamponade causing obstructive cardiogenic shock, timely diagnosis is crucial, with appropriate treatment through pericardial fluid drainage and simultaneous volume replacement. In cases of suspected chylopericardium, early recognition of lymphatic fluid leakage allows prompt treatment with an MCT-rich diet, possibly enhancing the effectiveness of conservative treatment without the need for surgery, leading to fluid resorption and reducing the risk of recurrence, along with management through administration of somatostatin analogues for a short period. Conservative management is effective in most cases and only if there is no valid clinical-instrumental response surgical intervention is recommended. Lastly, considering the possibility of recurrence and complications such as the development of acute or constrictive pericarditis in the long term, regular follow-up over time is important

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