




## CASE REPORT

# Abdominal aortic thrombus formation in a neonate with an interrupted aortic arch

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## Abstract

We note the risk of paradoxical embolism in patients with congenital heart defects with a right-to-left shunt. These patients should be managed to ensure that abdominal aortic thrombi are not overlooked when their clinical conditions change.

## KEYWORDS

abdominal aortic thrombus, congenital cardiac defects, interrupted aortic arch, neonate, paradoxical embolism

## 1 | INTRODUCTION

We report a case of abdominal aortic thrombus in a neonate with an interrupted aortic arch and ventricular septal defect. The thrombus moved from the main pulmonary artery to the abdominal aorta. In children with right-to-left shunt, venous thrombi can flow into the systemic circulation, depending on cardiovascular system hemodynamics.

Thrombus formation is one of the most frequent complications related to central venous catheterization,<sup>1</sup> which is

often mandated for the management of severely ill infants, including those with complex congenital heart defects. The venous thrombus may drift across an intracardiac right-to-left shunt and cause an embolism in the systemic circulation, known as a paradoxical embolism.<sup>2,3</sup> While embolisms occurring in cerebral arteries are relatively common, there has been no previous report of a paradoxical embolism occurring in the abdominal aorta in neonates. Here, we present a neonatal case of abdominal paradoxical embolism, in which the venous thrombus deviated from the venous stream due to a

congenital cardiac defect. We also reviewed previous cases of neonatal paradoxical embolism and discussed the key to its diagnosis.

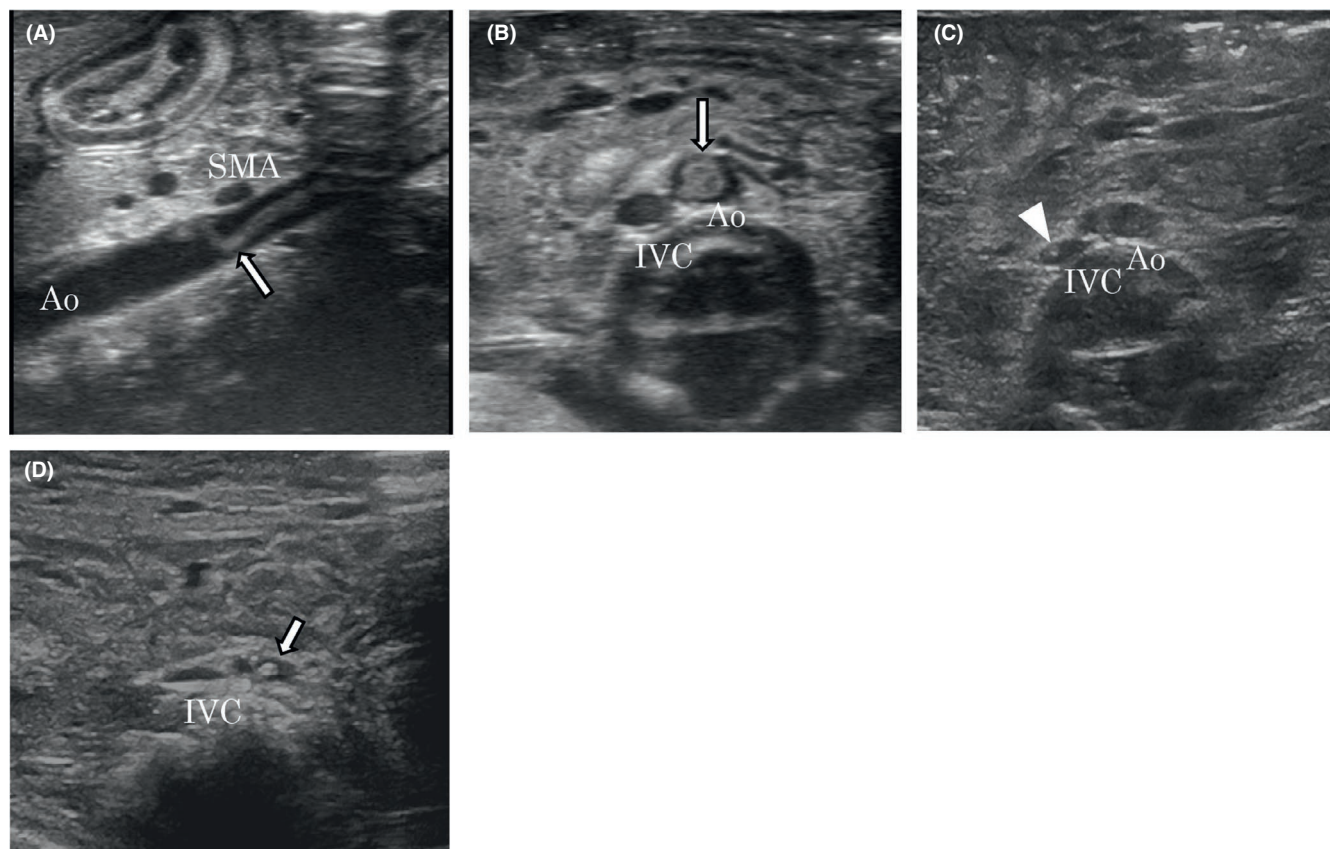
## 1.1 | Case History/Examination

A female infant was born at 34 weeks of gestation weighing 1,742 g. Aortic arch interruption was suspected prenatally, and she was delivered by cesarean section due to nonreassuring fetal status. Postnatal cardiac sonography confirmed the diagnosis of interrupted aortic arch with a ventricular septum defect. A peripherally inserted percutaneous central catheter, Argyle™ 28G single lumen, was immediately placed into her right femoral vein, with the tip placed in the inferior vena cava, distal to the renal vein. Prostaglandin E1 was initiated 2 hours after birth to maintain the “pulmonary-ductus descending aorta trunk.” Subsequently, she was transferred to our neonatal intensive care unit (NICU) 14 hours after birth. Overlapping fingers and hypoplasia of the corpus callosum were noted, but G-band karyotyping analysis showed normal results.

## 1.2 | Differential diagnosis, investigations, and treatment

Bilateral pulmonary artery banding was performed 8 days after birth to prevent pulmonary over-circulation. The postoperative course was initially uneventful. On postoperative day 7, her right lower limb was found to be swollen and pale in color. Removal of the previously inserted percutaneous central venous catheter ameliorated these symptoms. The following day, her oxygen saturation dropped sharply without any known reason, followed by a rapid drop in heart rate to 50 bpm. She was resuscitated after 12 minutes of cardiopulmonary resuscitation. Despite this significant collapse, her hemodynamics remained completely stable afterward without inotropic support. However, she passed mildly bloody stools shortly after the event.

To investigate the cause of hematochezia, abdominal ultrasound was performed, which revealed a large, 30-mm long thrombus floating in the abdominal aorta (Figure 1A, B). Blood flow was preserved in the bilateral iliac arteries, celiac trunk, and superior mesenteric artery. A 1-mm thrombus was



**FIGURE 1** Abdominal ultrasound images from a patient with continuous hematochezia. A: Sagittal view of the middle abdomen. A large thrombus (arrow) is attached to the orifice of the celiac trunk and extends to the bifurcation of the iliac arteries. B: Axial view of the middle abdomen. The thrombus almost completely occupies the aorta. C: Axial view of the abdomen at a lower level than that in panel B. A small 1-mm thrombus (white arrowhead) is detected in the IVC. D: One month after heparin treatment, the large thrombus is reduced to a thrombus 1 mm in diameter and is fixed on the intima of the aorta. Hyperechogenic structure suggests the thrombus organized. Ao: aorta, SMA: superior mesenteric artery, and IVC: inferior vena cava

also noticed in the inferior vena cava (Figure 1C). Systemic heparinization was commenced targeting an activated partial thromboplastin time of 55 sec. Table 1 displays the patient's blood test results recorded around the day of the hemodynamic collapse event. There were no findings suggestive of infection or dehydration, and her cardiac function was normal. However, during postnatal days 5-8, there was a sudden decrease in fibrinogen levels and an increase in the levels of fibrin degradation products.

### 1.3 | Outcome and follow-up

Repeated ultrasound showed a gradual regression of the thrombus over time. One month later, the aortic thrombus had reduced to a 1-mm immobile thrombus (Figure 1D). Computed tomography with contrast media 8 months after birth revealed no signs of abdominal thrombus or visceral infarction.

## 2 | DISCUSSION

An international multicenter registry reported the incidence of venous thromboembolism as 2.4/1000 among neonates admitted to the NICU,<sup>4</sup> for which the greatest risk factor is central venous catheterization. However, the cause of occult

thrombosis is not fully understood. A previous study suggested the prevalence of occult thrombus formation related to umbilical vein catheterization to be as high as 20-65%.<sup>5</sup> Extra attention should be paid to thromboembolism while managing neonates with central venous catheters. The most effective strategy for preventing catheter-related thrombus formation is early removal of the catheter.<sup>1</sup> However, long-term central venous catheterization is often required in the management of neonates with complex congenital heart disease, especially for those requiring continuous prostaglandin infusion. Moreover, these patients could be exposed to other thrombotic risks, including valvular regurgitation, heart failure, infection, and surgical invasion.<sup>4,6</sup>

In our case, the patient had an aortic arch interruption and therefore required continuous central venous catheterization for prostaglandin E1 infusion. We assumed that the venous thrombus had flowed into and blocked a main pulmonary artery, which caused a temporal hemodynamic collapse on postoperative day 8. Rapid recovery after cardiopulmonary resuscitation indicated that the thrombus had temporarily blocked a main pulmonary artery but immediately drifted across an intracardiac right-to-left shunt and flowed into the abdominal aorta. In addition, the thrombus might not have moved to the bilateral pulmonary artery because of banding surgery.

Paradoxical embolism is suspected when an arterial embolism and a venous thrombus (including pulmonary embolism) are noted concurrently in the presence of an intracardiac right-to-left shunt.<sup>2</sup> Our patient fulfilled these requirements. Although we cannot exclude the possibility of simultaneous thrombus formation in the veins and arteries, the patient had not been exposed to known risk factors for arterial thrombi, such as arterial catheterization, sepsis, and coagulation disorders.<sup>7</sup>

To our knowledge, only four cases of neonatal paradoxical embolism have been previously reported. The characteristics of these cases, in addition to those of our case, are summarized in Table 2.<sup>5,8-10</sup> Our case was unique in that the thrombus was observed in the abdominal aorta. In general, cardiac thrombi are 10 times more likely to be associated with a cerebral embolism<sup>11</sup> than other parenchymatous or limb embolisms. Indeed, a paradoxical thrombus in neonates occurred in the cerebral or coronary arteries in previous cases, while in our case, the thrombus occurred in the descending aorta. Since our patient had an aortic arch interruption and ventricular septum defect, as well as postbilateral pulmonary artery banding, it was assumed that thrombus movement to the abdominal aorta was facilitated by the blood stream (Figure 2).

Symptoms of abdominal thrombi are commonly nonspecific, such as abdominal symptoms (16%) and respiratory disturbances (14%).<sup>7</sup> Therefore, physicians should pay attention to these nonspecific symptoms and consider the possibility of an embolism associated with the structure of the

**TABLE 1** The patient's blood test results recorded around the day of the hemodynamic collapse event

White blood cell count	9400 / $\mu$ L
Red blood cell count	442 $\times 10^4$ / $\mu$ L
Hemoglobin	15.8 g/dL
Platelet count	40 $\times 10^4$ / $\mu$ L
Aspartate aminotransferase	36 U/L
Alanine aminotransferase	2 U/L
Lactate dehydrogenase	693 U/L
Creatine kinase	480 U/L
C-reactive protein	0.94 mg/dL
Serum creatinine	0.64 mg/dL
Anti-CL $\beta$ 2GPI complex antibody	<0.7 U/mL
Lupus anticoagulant	1.0
Activated partial thromboplastin time	29.3 sec
Prothrombin time	21.0 sec
Fibrinogen	310.0 mg/dL
Antithrombin activity	86.0%
Fibrin/fibrinogen degradation products	11.4 $\mu$ g/mL
D-dimer	4.9 $\mu$ g/mL
Protein S	81%
Protein C	68%

TABLE 2 Review of neonatal paradoxical embolism cases previously reported

No	Reported year	sex	Weight (weeks)	pre-existing comorbidities	Day*	origin of the thrombus	shunt	location of the thrombus	symptoms	discharge status
1	1979	f	3000 (44w)	bilateral pneumothorax and meconium aspiration	2	tip of the central venous catheter (umbilical vein)	PFO	left middle cerebral artery	seizure	died
2	2001	f	4040 (term)	shoulder dystocia	0	ductus venosus (no catheter)	PFO	left main coronary artery	sudden circulatory collapse	died
3	2004	m	950 (27w)	respiratory distress syndrome	17	tip of the central venous catheter (left lower leg)	PFO	left middle cerebral artery	sudden respiratory arrest, seizure	survived, paresis of the right arm
4	2016	f	n.d. (30w)	none	1	tip of the central venous catheter (right lower leg)	PFO	pulmonary artery, aortic arch, cerebral artery	sudden circulatory deterioration	survived, psychomotor retardation
5	our case	f	1742 (34w)	interrupted aortic arch, ventricular septal defect	12	tip of the central venous catheter (right lower leg)	PDA	abdominal aorta	sudden circulatory deterioration, bloody stool	survived, psychomotor retardation

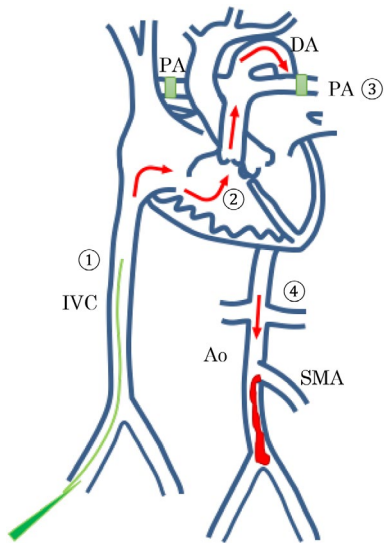


FIGURE 2 Schema of the suspected mechanism causing a thrombus in the abdominal aorta in this case. ① PICC-related thrombus is formed in the venous system. ② The clot travels into the heart and is trapped in the main PA; ③ the clot migrates into the artery via the DA. ④ Bilateral PA banding suppresses PA flow. ④ The thrombus is fixed on the orifice of the SMA. PICC: peripherally inserted central catheter, PA: pulmonary artery, Ao: aorta, SMA: superior mesenteric artery, IVC: inferior vena cava, and DA: ductus arteriosus

patient's cardiovascular system to predict its location. In our case, we were able to detect the thrombus because of the passage of bloody stools. A sudden decrease in fibrinogen levels and an increase in the levels of fibrin degradation products might also be indicative of a thrombus<sup>12</sup>; therefore, the potential presence of thrombi should be investigated in these situations.

3 | CONCLUSIONS

We reported the first case of a paradoxical embolism in the abdominal aorta occurring in a newborn with an aortic arch interruption and a ventricular septum defect. In this case, a catheter-related thrombus in the inferior vena cava drifted across an intracardiac right-to-left shunt and settled in the abdominal aorta. Based on this, we note the risk of paradoxical embolism in patients with congenital heart defects with a right-to-left shunt. These patients should be carefully managed to ensure that abdominal aortic thrombi are not overlooked when their clinical conditions change.

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## CONFLICT OF INTEREST

All authors have no conflicts of interest.

## AUTHOR CONTRIBUTIONS

YY, TK, and KI: participated in the conceptualization and writing of the manuscript. YY, YN, TI, NI, HK, KY, MT, TM, HH, and HT: helped draft the manuscript. All authors read and approved the final manuscript.

## ETHICAL STATEMENT

Written informed consent was obtained from the patient's parent for the publication of the case report.

## DATA AVAILABILITY STATEMENT

The data of the study are available from the corresponding author upon reasonable request.

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