Images in Pediatric and Congenital Heart Disease

Prominent Angulation of the Inferior Vena Cava in a Male Patient with Repaired Omphalocele: Significance for Interventional Cardiologists

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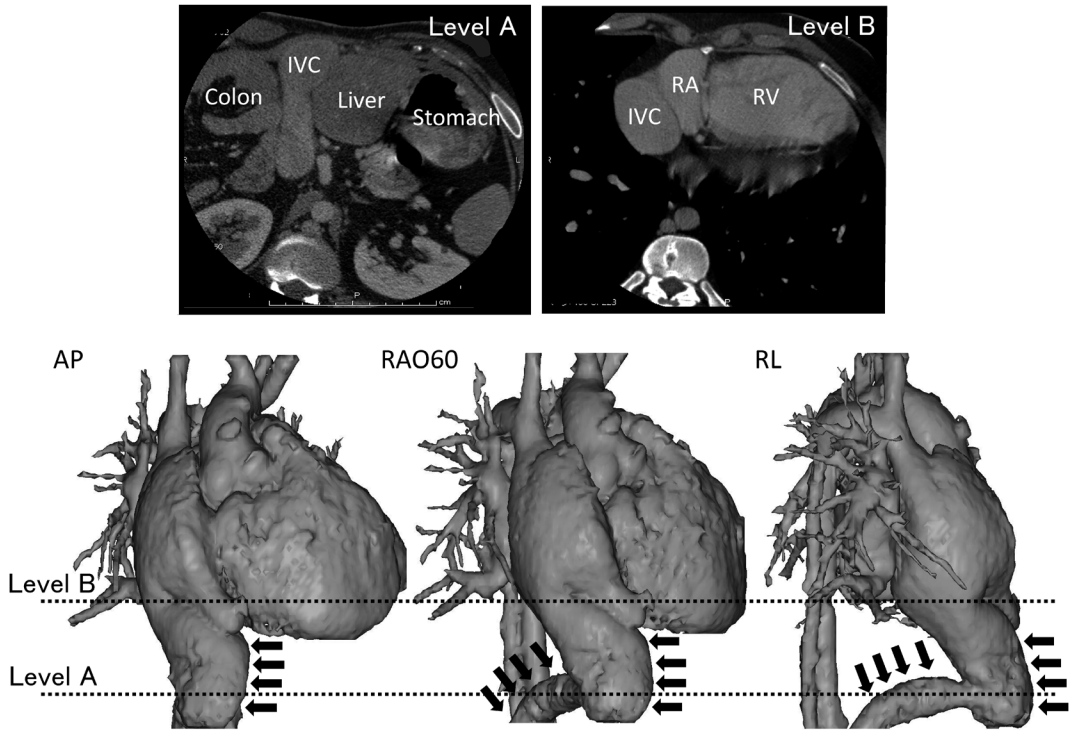

Fig. 1 Enhanced cardiac computed tomography (CT) (top) and three-dimensional reconstruction of CT angiography images (bottom)

At the subdiaphragmatic portion (Level A), the inferior vena cava (arrows) turned anteriorly and ran superiorly. It was connected to the anterolateral portion of the right atrium (Level B). IVC, inferior vena cava; RA, right atrium; RV, right ventricle; AP, anterior-posterior view; RAO60, 60-degree anterior oblique view; RL, right lateral view.

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A 21-year-old man was referred for electrophysiological evaluation and catheter ablation of recurrent atrial tachycardia that was refractory to multiple antiarrhythmics. He had tetralogy of Fallot and omphalocele repairs in childhood.

At the beginning of the procedure, we attempted catheter entry via the femoral veins. At the subdiaphragmatic level, however, vascular obstruction was suspected. Venography revealed marked angulation of the inferior vena cava (IVC) (Fig. 1). Because of significant limitations in the number of insertable catheters and inoperability by the femoral venous approach, we switched to right internal jugular venous access. Using a three dimensional electroanatomical mapping system, the patient's induced clinical arrhythmia was diagnosed as common atrial flutter. Linear ablation was performed at the cavo-tricuspid isthmus. However, it was difficult to confirm blocking lines because of the limited number of diagnostic electrode catheters, and his arrhythmia was not inducible at the end of the session. He remained recurrence-free during 3 years of follow-up.

Omphalocele is an anterior abdominal wall defect at the base of the umbilical cord resulting in herniation of the abdominal contents. It occurs in approximately 1 in 5000 live births.[1] The incidence of associated structural anomalies generally ranges from 35 to 70%, with cardiac defects occurring in up to 50% of cases. The survival rate after radical omphalocele surgery is approximately 80%.[2] Various abnormalities of the systemic veins associated with omphalocele have been reported, including an abnormally angulated, obstructed, or interrupted IVC with continuation.[3] The frequency of these vessel anomalies is uncertain, because patients remain asymptomatic without blood flow disturbance.

Systemic venous anomalies and omphalocele develop coincidently during the embryonic period (gestational weeks 6–11). Most of these vessel abnormalities are congenital, but some are postoperative sequelae due to compression of the veins.[1]

Recently, the number of cardiac catheterizations and interventions in adult patients with congenital heart disease has been increasing.

We cardiac interventionists should pay special attention to such patients to ensure blood access. Echocardiography may be helpful to detect these vessel anomalies in early childhood.

In patients with repaired omphalocele, confirmation of the anatomy of abdominal vessels before cardiac catheterizations is very important.

References

1) Marshall J, Salemi JL, Tanner JP, et al: Prevalence, correlates, and outcomes of omphalocelein the United States, 1995–2005. Obstet Gynecol 2015; **126**: 284–293
2) Corey KM, Hornik CP, Laughon MM, et al: Frequency of anomalies and hospital outcomes in infants with gastroschisis and omphalocele. Early Hum Dev 2014; **90**: 421–424
3) Mlczoch E, Carvalho JS: Interrupted inferior vena cava in fetuses with omphalocele: Case series of fetuses referred for fetal echocardiography and review of the literature. Early Hum Dev 2015; **91**: 1–6