Patients in our study with heterotaxy and congenital heart disease (CHD) showed high postoperative morbidity/mortality, with some having respiratory complications.² Although this respiratory phenotype is often attributed to the CHD, airway mucus clearance and left-right patterning, abnormal in heterotaxy syndrome, both require motile ciliary function. Thus airway ciliary dysfunction (CD) similar to that of primary ciliary dyskinesia (PCD) may have relevance for increased respiratory complications seen in patients with heterotaxy. To better characterize the phenotype of a broad spectrum of patients with heterotaxy, 43 patients with CHD were assessed for airway CD. We used videomicroscopy to look for abnormal ciliary motion in nasal tissue and nasal nitric oxide noninvasive testing (typically low with PCD) to assess for CD. A remarkable 42% (18/43) of patients exhibited CD, with one third in the severe PCD range. Patients greater than 6 years old with CD showed increased respiratory disease similar to that seen in PCD.³ Our studies suggest patients with CHD who have heterotaxy have substantial risk of CD and respiratory disease that complicates their prognosis.

The phenotype-genotype-prognosis correlation is also important in that further research may allow future patients and families to correlate gene mutations with phenotype and prognosis. Now families face the very high recurrence risk ratio of 79.1 for heterotaxia (95% confidence interval, 32.9-190).⁴ Further study of our patients with heterotaxy who have CD through next-generation sequencing of all 14 known PCD genes in 13 heterotaxy patients with CD identified 10 novel coding variants in 6 PCD genes. Recently, some CD gene mutations have become commercially available. The paradigm of phenotype-genotype-prognosis is beneficial to promote translational research to the patient, family, and medical care team. Our research suggests that patients with CHD and heterotaxy will

be at higher perioperative risk if they have CD.³ These findings indicate that patients with CHD and heterotaxy may benefit from preoperative screening for CD. Recognition of specific surgical risks factors in genetic syndromes is leading to specific diagnostic protocols that improve phenotyping or predictive genotyping.¹ To add "therapy" to the paradigm, further studies are needed to evaluate whether perioperative protocols improving airway ciliary motility and enhancing mucus clearance may reduce respiratory complications and improve postoperative outcome for patients with CHD who have heterotaxy and CD.

Linda Leatherbury, MD^a

Cecilia W. Lo, PhD^b

Richard Jonas, MD^a

^aChildren's National Heart Institute
Children's National Medical Center
Washington, DC

^bDepartment of Developmental
Biology

University of Pittsburgh School of
Medicine
Pittsburgh, Pa

References

1. Formigari R, Michielon G, Digilio MC, Piacentini G, Carotti A, Giardini A, et al. Genetic syndromes and congenital heart defects: how is surgical management affected? *Eur J Cardiothorac Surg*. 2009;35:606-14.
2. Swisher M, Jonas R, Tian X, Lee ES, Lo CW, Leatherbury L. Increased postoperative and respiratory complications in patients with congenital heart disease associated with heterotaxy. *J Thorac Cardiovasc Surg*. 2011;141:637-44.
3. Nakhleh N, Swisher M, Francis R, Giese R, Chatterjee B, Connelly P, et al. Low nasal nitric oxide and ciliary dysmotility in patients with congenital heart disease and heterotaxy. *Am J Respir Crit Care Med*. 2009;179:A1223.
4. Oyen N, Poulsen G, Boyd HA, Wohlfahrt J, Jensen PKA, Melbye M. Recurrence of congenital heart defects in families. *Circulation*. 2009;120:295-301.

doi:10.1016/j.jtcvs.2011.11.006

LATE EROSION OF AN AMPLATZER SEPTAL OCCLUDER DEVICE

To the Editor:

We read with interest the article "Late erosion of an Amplatzer septal

occluder device 6 years after placement" by Taggart, Dearani, and Hagler¹ in the *Journal of Thoracic and Cardiovascular Surgery*.

It is assumed that cardiac perforation is a rare complication following transcatheter device closure of an atrial septal defect (ASD) or patent foramen ovale (PFO). We, too, had a patient who had similar complications of perforation 9 months after closure of an ASD with a BioSTAR septal occluder (NMT Medical, Boston, Mass).² Another patient came to us 3 years after ASD closure with a Solysafe device (Swissimplant AG, Solothurn, Switzerland) with complications of device fracture and embolization into the pulmonary artery. Our patients warranted emergency surgical exploration for device explantation, repair of the erosion, along with pericardial patch closure of the residual septal defect. Although innovative absorbable closure devices that gradually biodegrade after implantation have theoretically decreased the long-term complications, it must be borne in mind that these devices are not totally bioabsorbed. The nonabsorbable components, such as the metal struts, may create long-term complications by eroding into the adjacent tissues.

This case clearly illustrates that inasmuch as tissue erosion, and other complications such as strut fracture, may occur at any time starting from 72 hours to 6 years after implantation, echocardiographic follow-up is needed, not just in the early stages, but also in the long-term. We also believe that clearer guidelines and recommendations may help prevent the complications associated with septal occluder devices.

Although cardiac perforation may be "rare," it is a potentially "serious" complication after transcatheter device closure procedures. These patients arrived at the hospital in time to benefit from immediate diagnosis and emergency surgical treatment of their complications. The question we need to ask ourselves and also

for our patients is whether the main “advantage” of transcatheter device closure, namely, its minimally invasive nature, is really worth taking the risk of having a life-threatening complication at a later date, requiring emergency surgery with eventual surgical closure of the septal defect. In contrast, primary surgical closure of an ASD/PFO through a minithoracotomy/ministernotomy is a safe and effective minimally invasive procedure, without the risk of erosion.

It is possible that some patients may have had only minor complications after transcatheter device closure, without the severe clinical complications presented in our case reports. Furthermore, how many centers are really reporting all the postimplantation complications? How many patients are being followed up to assess long-term complications?

The other question we should ask ourselves is, “Are we really only seeing the tip of the iceberg?” Moreover, is even the tip of the iceberg really much bigger than what is being presented?

Sanjay Cherian, MD, PhD, FRCS
Afksendiyos Kalangos,
MD, PhD, FETCS, Dsci(Hon)
Mustafa Cikirikcioglu,
MD, PhD, FETCS
Department of Cardiovascular
Surgery
University Hospital of Geneva
Geneva, Switzerland

References

1. Taggart NW, Dearani JA, Hagler DJ. Late erosion of an Amplatzer septal occluder device 6 years after placement. *J Thorac Cardiovasc Surg.* 2011;142:221-2.
2. Cikirikcioglu M, Cherian S, Lerch R, Licker M, Roffi M, Bonvini M, et al. Late tamponade secondary to aortic root perforation by BioSTAR septal closure device. *Ann Thorac Surg.* 2011;91:604-6.

doi:10.1016/j.jtcvs.2011.10.087

Reply to the Editor:

We appreciate the conscientious input by Cherian, Kalangos, and

Cikirikcioglu regarding the risk of erosion associated with Amplatzer septal occluder devices (AGA Medical Corporation, Golden Valley, Minn). We likewise acknowledge that we do not know the true timeline of device erosions. This uncertainty may create some unease among cardiologists and surgeons, but it should not necessarily dissuade us from catheter-based treatment of atrial septal defects (ASDs). Certainly, one may speculate about icebergs, but there is little evidence of those here. The fact remains that device erosion is rare; current postmarket study of the Amplatzer septal occluder device placement may provide some insight into the true incidence of erosions.

Although the letter raises important questions to consider before selecting a transcatheter or surgical approach to treatment of ASDs, we disagree with the implication that erosion is an inherent risk of transcatheter ASD closure. The 2 devices mentioned in the letter (BioSTAR and Solysafe) both have stiff arms, which may perforate and have been removed from the US market. In addition, the Gore Helex device (W. L. Gore & Associates, Inc, Flagstaff, Ariz) has not been associated with any erosions and may represent a safe alternative to surgical intervention.

What we can all agree on is that discussions such as this one are vital to developing safer, less invasive treatment strategies for patients with congenital heart disease.

Nathaniel W. Taggart, MD
Donald J. Hagler, MD
Mayo Clinic
Rochester, Minn

doi:10.1016/j.jtcvs.2011.11.007

EN BLOC REMOVAL OF THE BRAIN AND SPINAL CORD FOR PERFUSION STUDIES

To the Editor:

We were a little puzzled during our reading of Al-Ali and colleagues'

recent article.¹ They mentioned that the brain and spinal cord were photographed and then carefully removed en bloc. Because the epidural space between spinal cord and vertebrae is small, spinal cord removed en bloc was not easy even if the spinal cords were fixed. Which methods and equipment were applied by Al-Ali and colleagues? In our experience, the vertebral column and cranium was decalcified first. Then craniotomy and laminectomy were easily performed to expose the brain and spinal cord in situ. The brain and spinal cord could then be removed en bloc.

In the study of Al-Ali and colleagues,¹ the length of filling in the anterior and posterior spinal arteries was also measured and then converted into a percentage of the total length of the cord. Sometimes, however, spinal arteries, especially the posterior spinal artery, are absent or partially absent (Figure 1). What did they do in such cases?

Al-Ali and colleagues reported measuring the length of the spinal cord from the brainstem to the conus medullaris.¹ In our experience, accurately identifying the initial point of the cauda equina is very hard without incision of the dura mater. How did Al-Ali and colleagues accurately distinguish between the conus medullaris and the cauda equina?

Zhong-Ke Lin, MMed
Quing Yu, MMed
Yong-Long Chi, BM
Department of Orthopaedic Surgery
Second Affiliated Hospital of
WenZhou Medical College
WenZhou, Zhejiang, China

Reference

1. Al-Ali S, Chen BS, Papali'i-Curtin AT, Timmings AR, Bergin C, Raudkivi P, et al. Adequacy of brain and spinal blood supply with antegrade cerebral perfusion in a rat model. *J Thorac Cardiovasc Surg.* 2011;141:1070-6.

doi:10.1016/j.jtcvs.2011.11.036