5. Filosso PL, Ruffini E, Oliaro A, Papalia E, Donati G, Rena O. Long-term survival of atypical bronchial carcinoids with liver metastases, treated with octreotide. *Eur J Cardiothorac Surg.* 2002;21:913-7.
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Reply to the Editor:

We read with interest the letter by Filosso from the University of Torino. When the patient came to our department, we had questions similar to those that Filosso posed.

In Japan, scintigraphy with indium In-111 pentetreotide (OctreoScan) is still only available for research purposes, not for clinical use. This is one of the reasons why we did not perform the scan before 2001, nor did we after the operation.

As Filosso mentioned, it is thought that bronchial carcinoid producing adrenocorticotrophic hormone is aggressive.¹ However, our patient showed a different clinical course, as we reported. More than 5 years passed between the onset of symptoms and her operation. Despite her long follow-up duration, no change in the size of mediastinal nodes was detected by serial examination until 2001. This may be the reason that her physician hesitated to consult us. In addition, the levels of adrenocorticotrophic hormone and cortisol had not become worse during her course.

With regard to the use of a mediastinoscope, we also believe that this procedure is a useful method to localize and diagnose a tumor. However, with this procedure, it is sometimes difficult to perform en bloc removal of the whole tumor. We were also afraid that piece by piece removal might cause an intraoperative crisis of Cushing syndrome. In addition, our previous studies^{2,3} showed that removal of lymph nodes could be completed under thoracoscopic surgery. Our data revealed that only 2% of lymph nodes in weight could not be removed when performing sequential thoracotomy just after thoracoscopic surgery. Regarding the region of pretracheal and paratracheal lymph nodes, no residual lymph nodes were detected in our studies. So in our case, we chose thoracoscopic surgery.

As mentioned by Filosso, we also believe that any pulmonary masses should be investigated in case of suspected ectopic adrenocorticotrophic hormone production. However, we did not detect any tiny lesions in either lung on thin-sliced computed tomographic images. In such case, should we perform bilateral thoracotomy? We informed our patient, and she chose thoracoscopic resection. We also told her and her family about the possible need for further surgery should a new lung mass shadow develop.

Octreotide is also a good alternative option for adjuvant therapy in cases of stage III carcinoid tumor after surgery to prevent recurrence.^{4,5} Before our patient's operation, she received octreotide treatment for about 1 year. She refused to continue the treatment, however, because of pain caused by subcutaneous injection. Now 2 years have passed since her operation. She is doing well without any signs of recurrence; the values of hormones are within normal ranges, and no abnormal mass has been detected by whole-body screening.

Finally, we appreciate Filosso's advice. He advised us to implement a follow-up plan including OctreoScan scintigraphy and possible octreotide treatment. Considering his advice, we will discuss her adjuvant therapy and follow-up plan with her physicians and perform careful and intensive follow-up.

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References

1. Shrager JB, Wright CD, Wain JC, Torchiana DF, Grillo HC, Mathisen DJ. Bronchopulmonary carcinoid tumors associated with Cushing's syndrome: a more aggressive variant of typical carcinoid. *J Thorac Cardiovasc Surg.* 1997;114:367-75.
2. Sagawa M, Sato M, Sakurada A, Matsumura Y, Endo C, Handa M, et al. A prospective trial of systematic nodal dissection for lung cancer by video-assisted thoracic surgery: can it be perfect? *Ann Thorac Surg.* 2002;74:900-4.
3. Kondo T, Sagawa M, Tanita T, Sato M, Ono S, Matsumura Y, et al. Is complete

systematic nodal dissection by thoracoscopic surgery possible? A prospective trial of video-assisted lobectomy for cancer of the right lung. *J Thorac Cardiovasc Surg.* 1998;116:651-2.

4. Filosso PL, Ruffini E, Oliaro A, Papalia E, Donati G, Rena O. Long-term survival of atypical bronchial carcinoids with liver metastasis treated with octreotide. *Eur J Cardiovasc Surg.* 2002;21:913-7.
5. Filosso PL, Rena O, Donati G, Casadio C, Ruffini E, Papalia E. Bronchial carcinoid tumors: surgical management and long-term outcome. *J Thorac Cardiovasc Surg.* 2002;123:303-9.

doi:10.1016/S0022-5223(03)00690-1

Spiral pattern: Universe, normal heart, and complex congenital defects

To the Editor:

In the fascinating article "Basic Science Review: The Helix and the Heart"¹ in a recent issue of *Journal*, Gerald Buckberg, supported by the anatomic studies of Torrent-Guasp and colleagues,² illustrated the spiral anatomy of the muscle bands and the helicoid shape of the myocardium. On the basis of recent observations on embryology and genetics, we have reached similar conclusions regarding the spiral pattern of the heart morphology in complex congenital defects.³

The normal heart has a clockwise spiral pattern of the outflow tracts and of the great arteries. The helicoid 3-dimensional movement driving to this condition, genetically determined in subjects with situs solitus, appears with the dextroventricular loop (right ventricle to the right, left ventricle to the left),⁴ progresses with the looping (anterior rotation of the right ventricle, posterior rotation of the left ventricle), and finishes with the spiral septations of the outflow tracts and the great arteries.⁵ Because the dextroventricular loop is the first recognized sign of asymmetry and lateralization of the body organs,⁴ the clockwise spiralization of the heart should be considered a specific pattern of chirality in vertebrates. In subjects with situs inversus, an incompletely understood genetic mechanism⁶ is responsible for a "mirror image" anatomy of the heart, with counterclockwise shape of the outflow tracts and great arteries. This condition appears with a

levoventricular loop (right ventricle to the left, left ventricle to the right), progresses with an inverted looping, and finishes with an inverted spiral septation of the outflow tracts and the great arteries. Therefore the spiral shape of the heart is the consequence of an asymmetric formation, and it derives from coordinated developmental mechanisms involving, in a 3-dimensional fashion, the segments of ventricles and great arteries. In contrast to patients with situs ambiguus the genetically determined anomalies of lateralization⁶ cause, at the cardiac level, an interruption of the helicoid pattern consisting in isomerism of the atria, random ventricular loop, and transposition of the great arteries. This is particularly frequent in patients with right isomerism (asplenia syndrome).³ To summarize these observations, the helicoid shape of the heart is genetically determined and may be clockwise in situs solitus, counterclockwise in situs inversus, and absent or interrupted in situs ambiguus.

The helicoid shape of the heart is interrupted in other types of complex congenital heart defects. In congenitally corrected transposition of the great arteries [S,L,L], there is situs solitus of the atria, but with a levoventricular loop and a transposition of the great arteries. The resulting anatomic pattern is neither clockwise nor counterclockwise. In this complex malformation, the helicoid shape of the heart is inverted at ventricular level and interrupted at the level of the great arteries. In complete transposition of the great arteries [S,D,D], despite the presence of situs solitus of the atria and dextroventricular loop, the parallel position of the great arteries shows the interruption of the spiral pattern of the heart. Experimental research on animals,⁷ clinical studies on familial recurrence,⁸ and results of human genetic mutations⁹ suggest that both these complex cardiac defects, congenitally corrected transposition of the great arteries and complete transposition of great arteries, could be included in the pathogenetic group of the defects of lateralization.

In conclusion, the concept of a spiral pattern of the heart may have important implications not only for the fields of myocardial morphology, pathophysiology, and cardiac surgery¹ but also for our understanding of the segmental morphogenetic

mechanisms of some complex congenital heart defects. There is no doubt that the helicoid shape of the heart recalls some structural aspects of plants and animals, and also patterns observed in the microcosm and macrocosm alike.^{1,3} The physic and genetic causes of these similarities will advance our efforts toward comprehension, if we will be able to maintain the attitude of the student.¹

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References

1. Buckberg GD. Basic science review: The helix and the heart. *J Thorac Cardiovasc Surg.* 2002;124:863-83.
2. Torrent-Guasp F, Ballester M, Buckberg GD, Carreras F, Flotats A, Carrio I, et al. Spatial orientation of the ventricular muscle band: physiologic contribution and surgical implications. *J Thorac Cardiovasc Surg.* 2001; 122:389-92.
3. Marino B. Transposition of the great arteries in asplenia and polysplenia phenotypes. *Am J Med Genet.* 2002;110:292-4.
4. Levine M, Johnson RL, Stern CD, Kehun M, Tabin C. A molecular pathway determining left-right asymmetry in chick embryogenesis. *Cell.* 1995;82:803-14.
5. de la Cruz MV, Sanchez-Gomez C, Arteaga MM, Arguello C. Experimental study of the development of the truncus and the conus in the chick embryo. *J Anat.* 1977;123:661-86.
6. Casey B. Two rights make a wrong: human left-right malformations. *Hum Mol Genet.* 1998;7:1565-71.
7. Yasui H, Morishima M, Nakazawa M, Aikawa E. Anomalous looping, atrioventricular cushion dysplasia, and unilateral ventricular hypoplasia in the mouse embryos with right isomerism induced by retinoic acid. *Anat Rec.* 1998;250:210-9.
8. Digilio MC, Casey W, Toscano A, Calabrò R, Pacileo G, Marasini M, et al. Complete transposition of the great arteries: patterns of congenital heart disease in familial recurrence. *Circulation.* 2001;104:2809-14.
9. Goldmuntz E, Bamford R, Karkera JD, de la Cruz J, Roessler E, Muenke M. *CFC1* mutations in patients with transposition of

the great arteries and double-outlet right ventricle. *Am J Hum Genet.* 2002;70:776-80.

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Cardiac transplantation in prisoners To the Editor:

I have just finished reading Drs McKnealy's and Sade's thoughtful editorial regarding cardiac transplantation in prisoners. A number of years ago I attended a transplant ethics conference. In the course of the meeting a case was presented of a young prisoner. He was suffering from heart failure and the issue arose as to whether he should be offered cardiac transplantation. Although I was only a conference attendee, the moderator recognized that I was the surgical director of our cardiac transplant program. The moderator asked me if such a patient would be offered cardiac transplantation at our institution. I stated that one of the cardiac transplant selection criteria was a stable psychosocial background. I did not believe that incarceration fulfilled that criterion. Thus, I informed the group that we would not offer such a patient a cardiac transplant.

Although I understand the ethical right that a prisoner has to this therapy and I am sensitive to the cost that such care imposes on the prison system, I have a more practical concern about offering transplantation to prisoners. Nelson Mandela aside, can we rely upon these individuals to participate in the close posttransplant follow-up necessary to ensure the success of this treatment? Can these patients be trusted to comply with their immunosuppressive drug regimen? I suspect that the answer to these questions is "no." The donor heart is a precious commodity and as such should be offered only to patients in whom there is a reasonable chance for long-term survival.

Thank you for providing us with a starting point for ongoing discussion of this very relevant dilemma.

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