
**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 adrenocorticotropic hormone is aggressive. 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 adrenocorticotropic 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 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 adrenocorticotropic hormone production. However, we did not detect any 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. 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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**Spiral pattern: Universe, normal heart, and complex congenital defects**

**To the Editor:**

In the fascinating article “Basic Science Review: The Helix and the Heart” in a recent issue of Journal, Gerald Buckberg, supported by the anatomic studies of Torrent-Guasp and colleagues, 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.

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), 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. Because the dextroventricular loop is the first recognized sign of asymmetry and lateralization of the body organs, 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 mechanism 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

**References**


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