sprung's disease, neuroblastoma, ganglioneuroblastoma, and lack of heart rate variability, and also with hypothalamic dysfunction. This suggests a primary defect of the stem serotonergic nerve cell or a neural crest migration abnormality. Bolande first coined the term "neurocrystopathy" to describe lesions resulting from aberrations in the early migration, growth and differentiation of neural crest cells. Neuroblasts from the neural crest migrate along side each of the developing spinal cord to give rise to the sympathetic ganglia. They also migrate ventrally to form the visceral autonomic ganglia, the chromaffin system, the Schwann cells investing all peripheral nerves, the leptomeninges and some of the central nervous system may also arise from the neural crest. Neuroblastoma, pheochromocytoma, ganglioneuroma, carcinoid tumors and Hirschsprung's disease may be related to the maldevelopment of these neural crest cells.

We have described a five-year-old girl with CCHS and mediastinal and adrenal ganglioneuromas. This is the first case report of an association between CCHS and a benign tumor of neural crest origin that we are aware of. Hunt et al. reporting three cases of CCHS, described one patient with multiple abdominal and thoracic ganglioneuroblastomas. Haddad et al. also described three cases of CCHS and Hirschsprung's disease, one of whom had multiple neural tumors. Bower et al. described a patient with CCHS, Hirschsprung's disease, recurrent diarrhea, and multiple abdominal and thoracic ganglioneuromas and neuroblastomas.

The reason for the reported association between CCHS and neural crest tumors is unclear. It is conceivable that as a result of early maldevelopment of the neural crest, there is an abnormality in the respiratory control pathway either in the chromaffin tissue of the carotid body chemoreceptors or in the afferent pathway (IX, X) nerves. It is possible that abnormally functioning chemoreceptors or their neural innervations could lead to sleep apnea or hyperventilation. Up to this time, MRI and CT scans have not revealed a specific brainstem lesion in primary CCHS.

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Accessory Thyroid Tissue in the Right Ventricle*

Elisabetta Doria, M.D.; Piergiuseppe Agostoni, M.D., F.C.C.P.; and Cesare Fiorentini, M.D.

In an asymptomatic 66-year-old woman presenting a systolic murmur in the pulmonic area, echocardiography evidenced a voluminous mass in the right ventricular outflow tract resembling a cardiac tumor. Histologic finding was that of aberrant normal thyroid. Although the occurrence of ectopic thyroid tissue in the heart is rare, it should be considered in the differential diagnosis whenever a mass is located at the level of the interventricular septum and encroaches on the right ventricular outflow tract. In these cases, a thyroid scintiscan may avoid an unnecessary surgical intervention.

(Chest 1989; 96:424-425)

Two dimensional echocardiography is the technique of choice for identification of intracardiac masses, since it recognizes the location of nearly 100 percent of thrombi and neoplasms. It has high sensitivity and specificity in determining morphology, spatial relationships and hemodynamic effects of intracardiac masses. The high risk of complications borne by catheterization in these diseases is an additional reason for restriction of diagnostic procedures to noninvasive methods. However, despite the progress in ultrasound techniques, the tissue histologic findings often remain uncertain. We describe the case of an intraventricular thyroid mass, mimicking a cardiac tumor.

CASE REPORT

A 66-year-old woman was admitted to our hospital with the echocardiographic diagnosis of intracardiac mass. The patient was asymptomatic. Cardiac auscultation revealed a loud midystolic murmur in the pulmonic area. Physical examination was otherwise normal. On chest roentgenogram, the only abnormality was a moderate increase of the heart transverse diameter. The ECG showed sinus rhythm and complete right bundle branch block. Routine laboratory test results, including thyroid hormone determination, were normal. Two dimensional echocardiography revealed normal left heart chambers and valves; right atrium and right ventricle were moderately enlarged; a voluminous intracavitary mass encroaching on the right ventricular outflow tract was visualized (Fig 1). The structure was approximately 4 cm long and 3 cm wide and extended from the interventricular septum to 2 cm behind the pulmonary valve; it appeared almost immobile, with smooth edges and was attached to the septum without a visible stalk; its gray scala appearance was dense and homogeneous. Anatomy of the pulmonary and tricuspid valves was normal. No other abnormal structure was visualized within the myocardium, in the pericardium, or in the caval veins. At Doppler echocardiography, moderate tricuspid regurgitation and turbulent flow in the pulmonic conus were present. Peak velocity of tricuspid regurgitant flow estimated a peak right ventricular systolic pressure around 60 mm Hg (Fig 1). The absence of peripheral vein thrombosis, the normal appearance of the right heart valves and walls, and the location of the mass in a high velocity flow region negated the interpretation of an intraventricular thrombus and suggested a neoplasm. Since

*From the Istituto di Cardiologia dell’ Universita degli Studi, Istituto Ricerche Cardiovascolari “G. Sisti”, Fondazione “F. Monzino”, Centro Ricerche Cardiovascolari del C.N.R., Milan, Italy.

Reprint requests: Dr. Doria, Istituto di Cardiologia, Universita di Milano, Via Fara 4, Milan, Italy 20138

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there was no evidence of a metastatic origin of the mass, we suspected a primitive cardiac tumor and agreed upon surgical intervention without further diagnostic investigations.

Intraoperative inspection confirmed the echocardiographic information. Due to the wide infiltration of the cardiac tissue extending from the interventricular septum to the pulmonary conus, the surgeon performed only a limited resection of the mass, which, at histology, showed the pattern of normal thyroid tissue. The postoperative course was uneventful. Postoperative echocardiogram revealed only a slight reduction of the mass size; however, peak right ventricular systolic pressure was reduced to 30 mm Hg. A thyroid scintiscan, performed after surgery, documented normal aspect and function of the thyroid lobes, as well as thyroid tissue at the heart level (Fig 2).

This case shows that the echocardiographic features of cardiac neoplasms may be shared by a rare, benign congenital malformation: cardiac thyroid. Cardiac thyroid is an extremely rare occurrence; to our knowledge only three cases of thyroid adenomas have been diagnosed by intraoperative biopsy.6 Our case is the first in vivo report of cardiac thyroid with normal tissue. At post-mortem examination, cardiac location of thyroid tissue has been detected in the pericardium, in the right ventricle, and in the interventricular septum.6 This peculiar location of thyroid tissue within the heart is related to the contact in the embryo between the developing ventral part of the heart and the pharynx.6

Since in vitro reports are very few, it is hard to suggest guidelines for the echocardiographic diagnosis of this disorder. Features shared by the three previously reported cases and the present one are the location at the right side of the interventricular septum and partial obstruction of the right ventricular outflow tract. We would suggest, therefore, a preoperative cardiac thyroid scintiscan in patients presenting with these echocardiographic features. The most immediate implication concerns a surgical decision which is mandatory for cardiac neoplasms and may be unnecessary for cardiac thyroid.

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