

## Unusual Tissue on the Septal Leaflet of the Tricuspid Valve Identified as Intracardiac Bronchogenic Cyst in a New-born

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### Abstract

A systolic murmur in a female newborn with no other clinical complaints indicated cardiologic evaluation. A ventricular septal defect and a hyperechogenic formation in the right atrium at the tricuspid valve septal leaflet without inflow obstruction were found. The suspected tricuspid valve aneurysm was at open heart surgery a thin-walled 6 × 8 mm large spherical formation protruding into the right atrium, containing thick yellowish liquid, sitting with a broad peduncle on the septal leaflet of the tricuspid valve. Microscopically, in the central part of the resected valve, there was a cystic space lined partly with ciliated columnar epithelium, partly with squamous nonkeratinizing epithelium. Underneath the epithelium was a layer of fibrous tissue rich in blood and lymphatic vessels, focally with myxoid changes and groups of smooth muscle fibres. The epithelial cells were positive for cytokeratin mixture AE 1/3, CEA, EMA and with nuclear TTF-1 expression, which lead to the diagnosis of intracardiac bronchogenic cyst. The presented case is the first published report of a bronchogenic cyst on the leaflet of the tricuspid valve in a neonate.

**Keywords:** Intracardiac bronchogenic cyst; Tricuspid valve mass; Neonate

### Introduction

Congenital heart diseases arise in 0.5% of new-born children and the percentage can even be higher in premature ones. These conditions may have a genetic reason or can be caused by environmental factors like infections or drugs [1]. Over 85% of congenital heart diseases are scattered in the following abnormalities of development: Shunts, defects causing obstructions to flow and miscellaneous defects. The remaining are primary and secondary tumours of the heart and cysts.

Bronchogenic cysts arise by abnormal folding or budding of the tracheal diverticulum in the 16<sup>th</sup> gestational week and originate from the ventral foregut forming the respiratory system. Thus, their typical location is the mediastinum or the lungs, atypically it appears in pericardium or the heart. They comprise 1% to 3% of all primary tumours of the heart and pericardium [2,3] and their prevalence in the population ranges from 0.0017% to 0.28% [4]. Most described cases of bronchogenic cysts were detected incidentally in adulthood, just two cases of an intracardiac cyst were reported in a child. Our finding of the lesion in a neonate is unique until now.

### Case Report

#### Clinical history

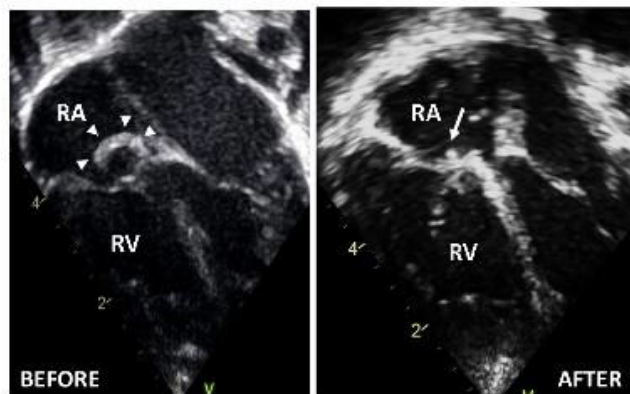
Postnatal screening of a female child born in the 40<sup>th</sup> week of gestation was in norm, a systolic murmur indicated cardiologic evaluation. A Ventricular Septal Defect (VSD) and an increased pulmonary flow were confirmed in the Children Cardiology Center. Another finding was a hyperechogenic formation in the right atrium,

which was described as redundant aneurysmal tissue of the tricuspid valve septal leaflet (Figure 1–BEFORE), causing no inflow obstruction to the right ventricle. The hyperechogenic formation was suspected as an aneurysm of the septal leaflet of the tricuspid valve. At the age of 7 months at open heart surgery the ventricular septal defect 15 × 12 mm was closed with a Dacron patch. The suspected aneurysm was a thin-walled 6 × 8 mm large spherical formation protruding into the right atrium, containing thick yellowish liquid, sitting with a broad peduncle on the septal leaflet. The lesion was resected and valvuloplasty of the tricuspid valve was performed.

At the last follow up control 1 year after surgery, the child was thriving and was cardially compensated. Echocardiography showed no residual ventricular shunt. Minor residual redundant tricuspid valve tissue at the site of aneurysm resection was found (Figure 1–AFTER), without any stenosis and with only trivial tricuspid valve insufficiency. At the time of the appointment no cardiologic therapy was indicated. Regular echocardiographic controls with the focus on the tricuspid valve were planned.

#### Histology and immunohistochemistry methods

The surgical specimen was fixed in 10% formalin, routinely processed in paraffin and 5 µm thick slices were cut and stained with hematoxylin and eosin. For immunohistochemistry, the slides after tissue epitopes demasking in Dako PT Link (Dako, Glostrup, Denmark) were incubated with antibodies against: Cytokeratin cocktail AE1/3, CEA, EMA, vimentin, CD10, CD30, CD31, CD34, CD45, CD56, smooth muscle actin, S100, NSE, NF, Calretinin, and TTF1. Positive binding was detected by En Vision FLEX/HRP system (Dako) and counterstained with hematoxylin.



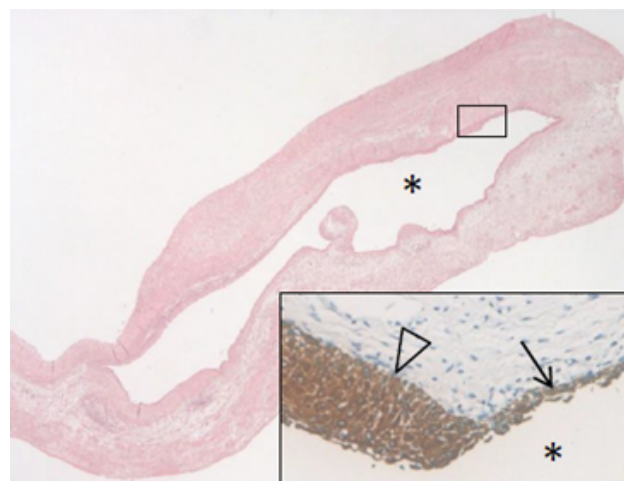
**Figure 1:** Echocardiographic findings before and after surgery. BEFORE surgery: Mobile cystic tumorous hyperechogenic tissue in the right atrium on the septal leaflet of the tricuspid valve, in diastole prolapsing into the right ventricle but without flow obstruction. 1 year AFTER surgery: Minimal residual abnormal tissue of the tricuspid valve leaflet (RA - Right Atrium, RV - Right Ventricle, arrow-abnormal tissue o the tricuspid valve).

### Pathology findings

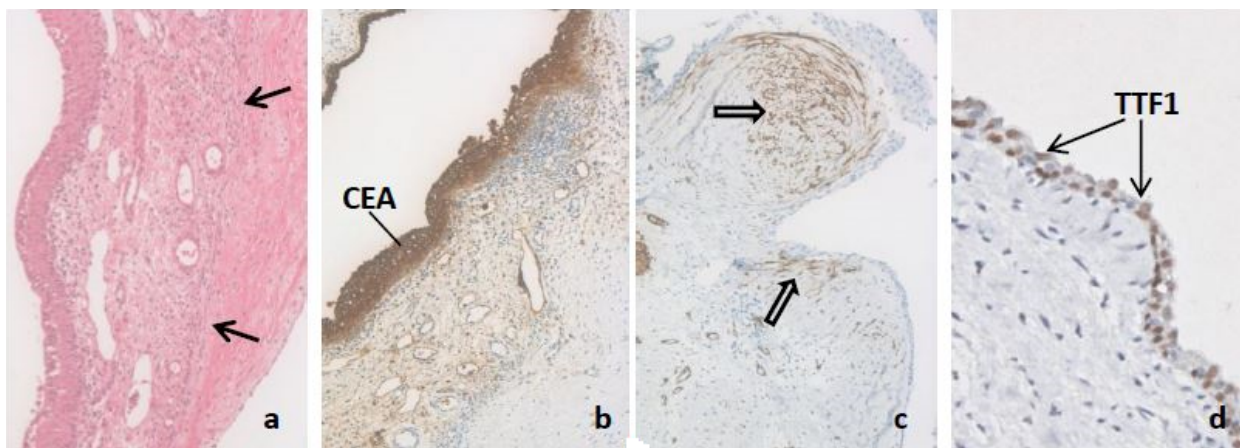
Surgical specimen consisted of elastic whitish valve material of semilunar shape 10 × 7 mm, 1 mm thick, without grossly identifiable focal changes.

Microscopically, in the central part of the resected valve, there was a cystic space lined partly with ciliated columnar epithelium, partly with squamous nonkeratinizing epithelium (Figure 2). Underneath the epithelium was a layer of fibrous tissue rich in blood and lymphatic

vessels, focally with myxoid changes and groups of smooth muscle fibers, clusters of lymphocytes or macrophages. Methenamine silver impregnation of collagen and staining of elastic fibers delineated the cystic formation from the valve tissue. Nerve and other types of tissues were not identified. Immunophenotype of the epithelium was CD30 negative, AE1/3 cytokeratin mixture positive, so with CEA, EMA and with nuclear TTF-1 positivity (Figure 3). Based on the above reported findings the diagnosis of intracardiac bronchogenic cyst was made.



**Figure 2:** Bronchogenic cyst of the septal leaflet of the tricuspid valve. The cystic space (\*) is lined with non-keratinising squamous epithelium (arrowhead) alternating with cuboid epithelium (arrow), stained with anti-cytokeratin antibodies cocktail in the figure insertion. Hematoxylin and eosin, 25x; AE1/3 antibodies, ABC-Px, 200x.



**Figure 3:** Bronchogenic cyst of the septal leaflet of the tricuspid valve. The wall of the cyst consists of richly vascularized connective tissue (arrow) with focal proliferations of smooth muscle cells (open arrow). The epithelial lining express carcinoembryonic antigen (CEA) and shows nuclear expression if thyroid transforming factor 1 (TTF1). **a.** Hematoxylin and eosin; **b.** CEA; **c.** Smooth muscle actin; 100x; **d.** TTF1; 200x.

## Discussion and Conclusion

To the best of our knowledge, our case is the first published report presenting a bronchogenic cyst on the leaflet of the tricuspid valve in a neonate. The only known case, in which an intracardiac mass attached to the tricuspid valve was found, was in a 2-year-old Nigerian boy [5]. Bronchogenic cyst is a benign cardiac tumor arising from an abnormality of the primitive foregut [3]. On day 21 after fertilization the developing heart tube gets into proximal vicinity to the ventral aspect of the foregut and the cysts will usually be localized in the pericardium and the mediastinum [6]. Rarely they are found intracardially, few intracardiac bronchogenic cysts were found in the right atrium [7], left atrium [8], interatrial septum [9], right ventricle [7] and in the left ventricle [10]. Bronchogenic cysts are often asymptomatic and found incidentally by murmurs, in other cases with variable clinical symptoms reflecting the location of the lesion, like dyspnea, chest pain, palpitations, severe cyanosis, systemic embolization or also as an AV-block [5,7,10].

Evaluation of these masses with echocardiography is very important since misdiagnoses of asymptomatic patients with a murmur are not infrequent [11]. In patients with no symptoms, surgical treatment is not mandatory unless infection or enlargement by mucin accumulation indicate removal for decreasing the risk of embolism, haemorrhage and rupture [7,12]. Magnetic resonance and computing tomography are additional non-invasive diagnostic methods helping to distinguish between solid and fluid components of the lesion [13].

The definite diagnosis is made by surgical excision. Microscopic finding elicited the suspicion of a mature teratoma of the tricuspid valve. Absence of tissues derived from multiple germ layers and the epithelium expression of TTF1 supported the bronchogenic origin. Bronchogenic cysts may resemble hydatid cysts and the differential diagnosis also includes tuberculosis, vascular malformations and neoplasms. To assess the exact diagnosis, tissue biopsy is necessary and complete extirpation is recommended to avoid recurrence and to contribute to a good prognosis [13].

As we could show in our case report, representing a unique rare finding of intracardiac bronchogenic cysts in a newborn, careful evaluation of the patient with focus directed in suspicion of the

uncommon is indicated to enable more target site diagnosis and adequate treatment.

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