# Life-Threatening Mediastinal Teratoma of Infant Requiring Emergency Surgical Removal

Nicole Piber[1] Wilko Weichert[2] Jürgen Hörer[1] Masamichi Ono[1]

[1] Deutsches Herzzentrum München des Freistaates Bayern, Munchen, Bayern, Germany
[2] Technical University of Munich Institute of Pathology and Pathological Anatomy, Munchen, Bayern, Germany

**Address for correspondence** Masamichi Ono, MD, PhD, Department for Congenital and Pediatric Heart Surgery, German Heart Center Munich, Lazarettstr.36, 80636 Munich, Germany (e-mail: Ono@dhm.mhn.de).

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## Abstract

**Background** Teratoma is a tumor derived from fetal germ cells with aberrant differentiation.

**Case Description** A 3-month-old infant with a mediastinal tumor was referred to our heart center. She presented with progressive dyspnea, cyanosis, and the need to be manually ventilated. The computed tomography scan displayed a huge tumor restricting the distal trachea including the bifurcation. An emergent operation was performed and the tumor was completely removed. Histological examination confirmed a mature teratoma.

**Conclusion** In such life-threatening situation, the early detection and the immediate operation are very important for the management of rapidly-progressing mediastinal teratomas compressing the respiratory tract.

**Keywords**
- mediastinal tumor
- teratoma
- infant

## Introduction

Teratoma is a tumor derived from fetal germ cells with aberrant differentiation. Following the common pathway of migration gonadal teratomas may be caused, whereas an abnormal migration might lead to extragonadal tumors.[1,2] Roughly, 5.4% of teratoma are found in the mediastinum mostly located in the anterior–superior area with a mortality of 18%.[2,3] Mediastinal teratoma presenting in the neonatal or early infantile age is uncommon.[4] They frequently present with severe respiratory distress requiring urgent surgery. We describe the case of an early infant with a mediastinal teratoma who presented with progressive dyspnea, cyanosis, and low oxygen saturation due to the airway compression, and needed emergent surgical removal of the tumor via a median sternotomy.

## Case Description

A 3-month-old female infant was referred to our heart center under the diagnosis of a mediastinal tumor for emergent surgical extirpation, eventually with the support of cardiopulmonary bypass or extracorporeal membrane oxygenation (ECMO). By admission, she presented with progressive dyspnea, cyanosis, and needed to be manually ventilated. The computed tomography (CT) scan displayed a tumor restricting the distal trachea including the bifurcation (►**Fig. 1A**). The ultra-sonogram showed hypo- and hyperdense areas and the size of $13 \times 9 \times 4$ cm with no infiltration in the pericardial tissue. Due to the life-threatening situation, the infant was transferred immediately from intensive care unit to the operation room. Surgeons were ready for emergency thoracotomy. Once unproblematic intubation was achieved,

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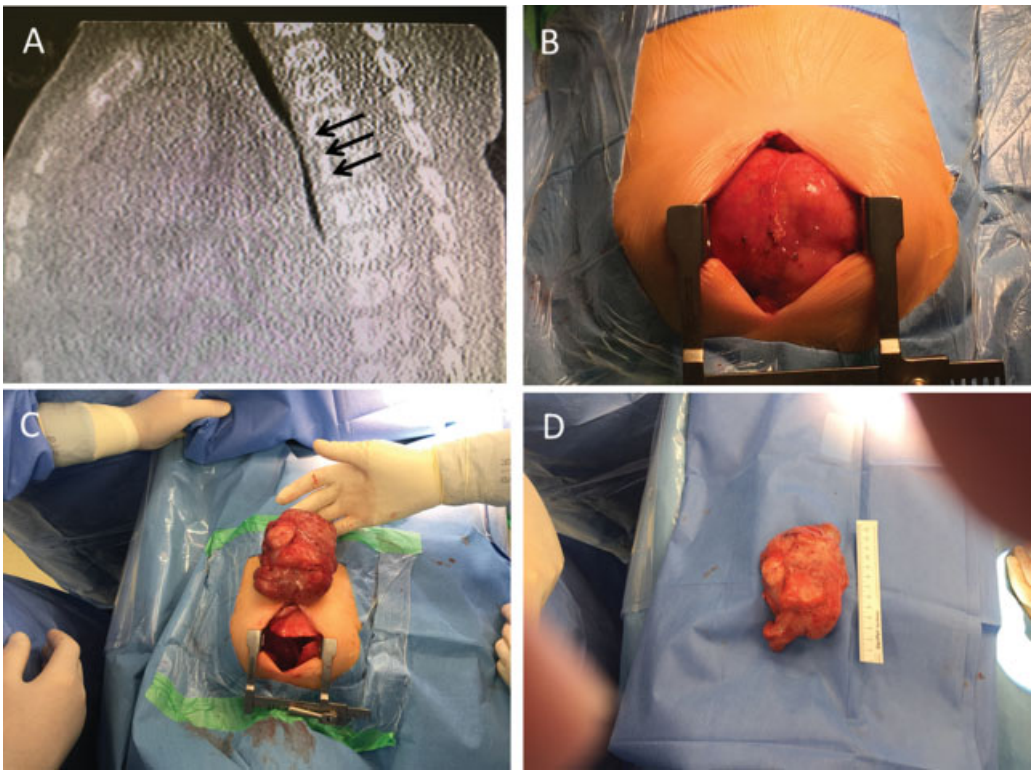


**Fig. 1** Preoperative and operative findings. (**A**) Preoperative computed tomography demonstrated a huge mediastinal tumor compressing the distal trachea (black Arrow). (**B**) After a median sternotomy, a large mass occupying the thorax was exposed. (**C**) The tumor was carefully dissected en bloc. (**D**) The resected tumor demonstrated cystic and solid areas with measurement of 12 × 7 cm.

median sternotomy was performed in usual fashion. The mass exposed itself directly substernal and extended from the thymic gland between pleural and pericardial tissue in both thoracic cavities (►**Fig. 1B**). Vascularization of the tumor was provided from a vessel deriving from the aortic arch and back to the innominate vein. No invasion of the tumor to the thoracic organs could be found. The tumor was carefully dissected en bloc (►**Fig. 1C** and **1D**). A misty effusion stood out in the left pleura that had to be drained. The postoperative recovery proceeded uneventfully. She was extubated on the first postoperative day and returned to the peripheral clinic on the second day postsurgery. At 15 months of follow-up, the patient was in good condition and showed no sign of recurrence.

## Histopathological Evaluation

The resected tumor measured 11.2 × 7.0 × 4.6 cm. The cut surface appeared brownish-yellow and featured mainly solid areas containing several cystic spaces (►**Fig. 2A**). By histology, a multitude of different differentiated tissue types were apparent. The arrangement appeared in a haphazardous fashion. Mature cartilage (►**Fig. 2B**), bone, fat, and muscle tissue were seen intermixed with brain parenchyma (►**Fig. 2C**), cutaneous adnexal structures as well as cysts lined by squamous and intestinal type epithelium containing goblet cells (►**Fig. 2D**). Primitive neuroectodermal tubules were not present. This finally led to the diagnosis of a mature teratoma (G0 according to Gonzalez-Crussi) featuring tissue from all three germinal layers.

## Discussion

Previous literatures described that germ cell tumors are present in one of 12,500 children under 15 years of age.[5] About 10 to 15% of premediastinal tumors in children turn out to be mediastinal teratomas that most likely arise from cells adjacent to the third and fourth brachial clefts.[6] Because of the rare location, the tumor is often misdiagnosed.[7] Mori et al described CT criteria on how to distinguish between teratoma and thymoma, a common misdiagnosis in terms of mediastinal tumors.[8] They demonstrated that teratomas are found to have a thicker wall and calcifications in a scattered lobular shape, whereas thymomas are lobulated and with no detectable wall between the surrounding structures. Calcification reminded of an eggshell.

The presentation of mediastinal teratomas in neonates and early infants is uncommon, and is sparsely documented.[4,9–12] Mediastinal teratomas presenting in the neonatal or early infantile period cause various degree of respiratory distress.[4] Therefore, urgent surgery is usually required. In severe cases, ECMO support is necessary to profound respiratory distress.[13–15] In the presenting case, the infant presented with severe respiratory distress because the tumor compressed the trachea. Emergent surgery immediately after the submission was performed under the

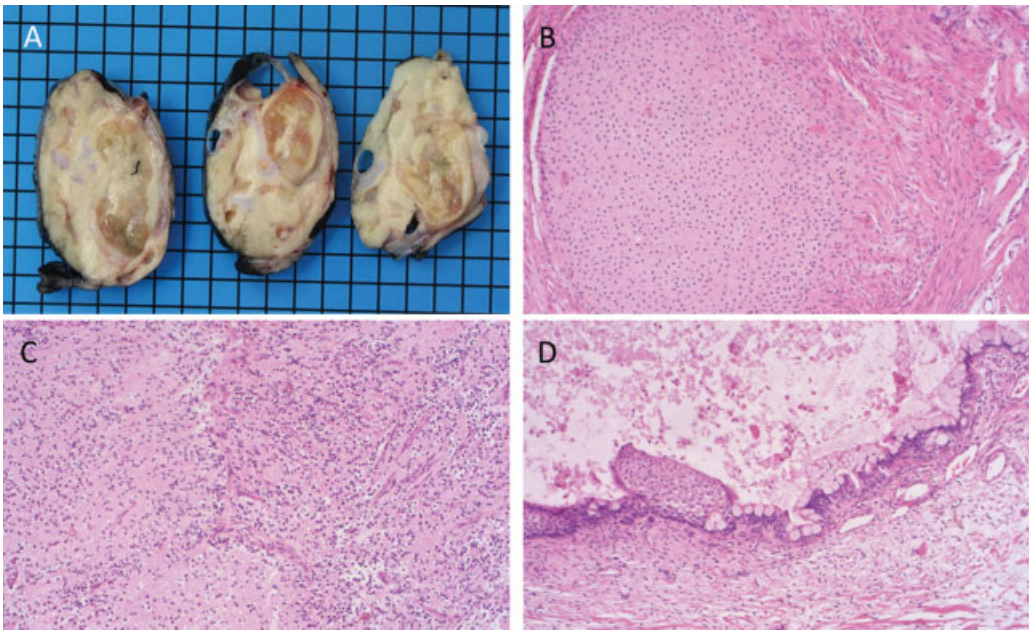


**Fig. 2** Histologic findings: mature teratoma of a newborn. (**A**) Cross-sections through the tumor with cystic and solid areas. The teratoma featured a multitude of haphazardly arranged differentiated tissue elements such as cartilage (**B**), cysts lined with squamous as well as cylindrical intestinal type epithelium containing brain parenchyma (**C**), and goblet cells (**D**).

stand-by situation for cardiopulmonary bypass and ECMO. Fortunately, intubation and ventilation were successfully performed without circulatory collapse and there was no need for cardiopulmonary support.

Complete surgical excision is essential for the treatment of mediastinal teratomas, and median sternotomy is the standard procedure especially for small patients, allowing good exposure and approachability of the tumor.[4] In the presenting case, no invasion of the tumor to the thoracic organs could be found, and the tumor was completely dissected. Histological examination confirmed the diagnosis of mature teratoma, and the patient showed no sign of recurrence at the last follow-up. The histological diagnosis and grading of tumor are crucial for planning the patient́s further management, and long-term follow-up is necessary for all patients to review the behavior of mediastinal teratoma.

In conclusion, we resected a huge mature mediastinal teratoma, which occupied the entire mediastinum and compressed the distal trachea including bifurcation in a 3-month-old female infant. In the life-threatening situation, surgical removal of the tumor was performed by pediatric cardiothoracic surgeons under the "stand-by" of cardiopulmonary bypass support which is thought to be inevitable for the successful perioperative management of the very ill infant.

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Conflict of Interest
There is no conflict exists. Author Nicole Piber declares that she has no conflict of interest. Author Wilko Weichert declares that he has no conflict of interest. Author Jürgen Hörer declares that he has no conflict of interest. Author Masamichi Ono declares that he has no conflict of interest.

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