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## Surgical approaches, anaesthetic management and outcome in pediatric superior mediastinal tumors

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## ABSTRACT

**Background:** Pediatric superior mediastinal tumors are a heterogeneous group of tumors with marked variation in pathology and extension. We reviewed our experience with different surgical approaches to tumors originating from or extending to superior mediastinum in pediatrics.

**Patients and methods:** The medical records of all patients who had undergone resection for superior mediastinal tumors in Children's Cancer Hospital – Egypt, between January 2008 to December 2015, were reviewed for demographic data, clinico-pathological features, radiologic findings, operative techniques and outcome.

**Results:** The study included 20 patients. Diagnosis included: germ cell tumors (n = 8), neuroblastoma (n = 4), soft tissue sarcoma (n = 3), thymolipoma (n = 2), infantile fibromatosis (n = 1), calcifying fibrous tumor (n = 1), and thymic carcinoma (n = 1). Tumor extension was divided into tumors extending unilaterally to one hemithorax (n = 9), tumors extending bilaterally to both hemithoraces (n = 4), and cervico thoracic junction tumors (n = 7). Extended lateral thoracotomy was used in 8 patients. Other approaches included trapdoor (n = 5), clamshell (n = 4), cervical approach (n = 2) and double level lateral thoracotomy (n = 1). There was no perioperative mortality, and postoperative morbidity was 20%. At the end of December 2016, 15 patients were alive free of disease, 5 patients developed local and/ or distant relapse.

**Conclusion:** Pediatric superior mediastinal tumors could be divided into 3 groups according to tumor extension. Each group has an optimum surgical approach that achieves the best exposure for adequate resection. However, further research is needed to confirm the conclusion as this was a descriptive study and the sample size was too small for valid statistical analysis.

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## Introduction

Primary tumors and cysts of the mediastinum in children and adults are uncommon [1–4]. In a collected series of primary mediastinal tumors and cysts, 25–49% of these lesions were malignant [1,2,4,5]. Pediatric primary mediastinal tumors are even less com-

mon, but the risk of malignancy is increased to approximately 75% [6,7]. The mediastinum is divided into compartments (superior, anterior, middle and posterior) and each compartment harbour characteristic tumors, however, they can occur in any of the mediastinal compartment due to the common occurrence of heterotopias of thymic tissue outside the anterior mediastinum [8]. Tumors arising from or extending to the superior mediastinal compartment in pediatrics are a heterogeneous group of tumors with marked variation in pathology, extension and response to chemotherapy. Due to the complexity of this compartment and variable tumor extension, different surgical approaches were

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described. Our objective was to review our experience with different surgical approaches to tumors originating from or extending to superior mediastinum in pediatrics.

## Patients and methods

### Patients selection

The study included all patients less than 18 years old at presentation who had undergone resection of tumors originating from or extending to the superior mediastinum between January 2008 and December 2015 at Children's Cancer Hospital, Egypt. All benign and malignant tumors that were a candidate for surgery either before or after chemotherapy were included. Cases proved to be lymphoma were excluded. During the same study period, there were 6 cases of cervicothoracic neuroblastoma encasing the major vessels in the thoracic inlet. All had >90% reduction in tumor size after chemotherapy but still encasing the major vessels at thoracic inlet. The decision was not to operate except in the case of tumor progression. All cases were followed up with no resection. All of them had a stationary disease.

### Study design

This was retrospective case review study. After scientific meeting accreditation committee (SMAC) approval, we reviewed the electronic medical records for the following: demographic data, clinicopathologic features, radiographic findings, operative techniques and outcome. All patients were followed up after surgery until the end of December 2016 or time to last contact with the patient for: perioperative complications, late postoperative complications; development of local recurrence or distant metastases and mortality. Patient characteristics were described in the overall cohort using count and percentage. Event-free survival (EFS) and overall survival (OS) were estimated using Kaplan–Meier analyses. Events were defined as relapse, progressive disease (PD), secondary malignancy or death. Survival outcome was presented  $\pm$  standard error. Analysis was performed using SPSS statistical package version 20.

### Preoperative evaluation and chemotherapy protocols

All patients had preoperative complete blood count, liver and renal functions and coagulation profile. Tumor markers (alpha-fetoprotein ( $\alpha$ FP) and beta human chorionic gonadotropin ( $\beta$  HCG)) was done if (GCT) was suspected radiologically. All patients had pre operative CT with intravenous contrast. CT angiography was done in cervicothoracic junction tumors encasing the vessels. A preoperative biopsy was done except if a benign diagnosis is concluded with certainty with imaging in a multidisciplinary team approach. After the biopsy, patients with malignant tumors received preoperative chemotherapy according to our hospital protocol as follow:

Patients with NB were intermediate risk patients and received Etoposide and Carboplatin (VP16/CARBO) alternating with cyclophosphamide, doxorubicin and vincristine (CADO), administered at 3-week intervals, with a total of 6 or 8 cycles. Patients with soft tissue sarcoma were on arm D protocol and received 4 cycles of chemotherapy before surgery and 4 cycles adjuvant with radiotherapy, in the form of 2 cycles IFX/ADR (ifosfamide/doxorubicin) and 2 cycles IFX/VP16 (ifosfamide/vepeside). One case of non-metastatic thymic carcinoma received 5 cycles of chemotherapy, 4 cycles preoperative and one cycle postoperative in the form of cisplatin, vepeside, cyclophosphamide and vincristine. While the 5 cases of germ cell tumors received 3 cycles of preoperative

chemotherapy in the form of cyclophosphamide, platinol, vepeside and bleomycin.

### Surgical approaches

**Posterolateral thoracotomy:** The patient is placed in a full lateral decubitus position with appropriate pressure point padding. The skin incision is started at the level of the anterior axillary line over the fifth intercostal space. It is curved around the tip of the scapula and continued posteriorly along a line between the medial aspect of the scapula and the spine. It is carried upwards to the level of T4. Anteriorly, the skin incision follows the rib outline. If an additional posterior extension is required, the anterior portion of the trapezius and rhomboid muscles can be divided. If an additional anterior extension is required, the skin incision is extended to the lateral edge of the sternum and the serratus anterior and pectoralis major muscles are divided. The mammary vessels are dissected and ligated in case partial sternotomy is needed.

**Trap-door:** The patient is placed in the supine position, the side of the anterior thoracotomy extension is elevated 30 degrees with a longitudinal roll placed beneath the scapula, and arms are tucked at the sides. A transverse incision is begun along the superior portion of the clavicle with descending median sternotomy through the midline sternum to the desired intercostal space to the anterior-axillary line.

**Clamshell:** The patient is placed in the supine position and the arms are extended. A curvilinear incision is made along the inframammary crease, extending from right to left anterior axillary lines, the mammary vessels are ligated and two Finochietto retractors are used to provide the retraction. The pleural reflections are incised to gain exposure to the mediastinal structures.

## Results

The study included 20 patients, 13 males and 7 females. The median age at diagnosis was 5 years (Range 2 months–16 years).

**Clinical presentation and preoperative evaluation:** There were 12 malignant and 8 benign tumors. The most common pathology was Germ cell tumor (GCT) followed by neuroblastoma. Shortness of breath was the commonest presentation (Table 1). Fifteen patients (75%) had initial radiologic guided biopsy. while 5 patients (25%) had surgery based on clinical and radiologic findings, thymolipoma (n = 2) and teratoma (n = 3). Eight patients had under-

**Table 1**  
patients and tumor characteristics.

Variable	Value		
Age at presentation	Median	5 y	
	Range	(2 M, 16 Years)	
Sex	Male	13	
	Female	7	
Presenting symptoms	Shortness of breath	6	
	Cough	5	
	Horner's syndrome	2	
	Pneumonia	2	
	Accidentally discovered	5	
Pathology	Benign/borderline	Mature teratoma	1
		Mature cytic teratoma	2
		Immature teratoma G 1	1
		Calcifying fibrous tumor	1
		Infantile fibromatosis	1
	Malignant	Thymolipoma	2
		Soft tissue sarcoma	3
		Malignant GCT	4
		NB	4
		Thymic carcinoma	1

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