Complex pediatric neoplasms: The role of congenital cardiothoracic surgery

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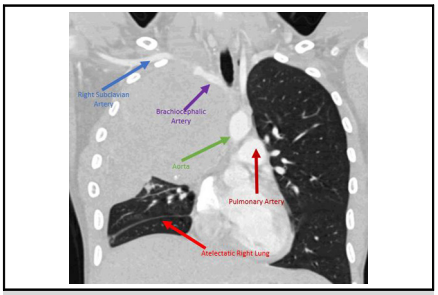
ABSTRACT

Background: Surgery for pediatric solid neoplasms is often complicated by local tumor invasion. Cardiac surgeons can provide expertise in the chest and facilitate potentially aggressive management of tumors invading vasculature, pericardiac, or diaphragmatic spaces. Here we present 4 complex cases.

Methods: This descriptive retrospective chart review study included 4 surgical patients with locally invasive solid tumors.

Results: Case 1: 16 × 15.5 × 11 cm right chest synovial sarcoma in a male patient status post–neoadjuvant chemoradiation. Imaging revealed invasion of the right-sided subclavian vein, subclavian artery, phrenic nerve, and vagus nerve. The surgical approach via hemi-clamshell allowed for R0 resection. Case 2: Resection of a 17.6 × 10.5 × 8.1 cm sclerosing epithelioid fibrosarcoma originating from the vertebral body but causing aortic arch, right and left pulmonary artery, tracheal, and esophageal displacement. The surgeons preserved nearly all thoracic anatomy despite extensive periaortic and posterior mediastinal dissection. Case 3: Synchronous removal of a 11.5 × 9 × 5.5 cm pleuropulmonary blastoma at the time of tetralogy of Fallot repair. Case 4: Resection of a 12 × 0.5 × 0.3 cm nonviable Wilms tumor traversing from the right renal vein to the level of the Eustachian valve. All patients were extubated in the operating room and had an uneventful hospital course, with length of stay ranging from 5 to 10 days.

Conclusions: Pediatric patients may present with locally advanced heterogenous neoplasms. The added anatomic familiarity with the mediastinum, thoracic hilum, and great vessels in particular ensured safe resection in all cases. Thus, cardiothoracic surgery consultation is valuable when managing complex thoracic oncologic tumor resection. (JTCVS Techniques 2025;30:137-43)



A right-sided partially calcified mass causing atelectasis and encasing vasculature.

CENTRAL MESSAGE

Congenital cardiothoracic surgeons may have expertise contributing to the safe completion of complex thoracic pediatric surgical oncology procedures.

PERSPECTIVE

Surgery for mediastinal tumors in children is rare and complex for those tumors surrounding and encasing critical structures. Complete resection may predispose patients to morbid complications. Including a congenital cardiothoracic surgeon in the multidisciplinary surgical team may improve the safety of these operations and enhance the likelihood of achieving complete resection.

Childhood solid tumors arising in the thorax and involving mediastinal structures are rare. For most nonhematologic solid tumors, the completeness of surgical resection has a significant impact on the prognosis and need for radiation therapy for local control.[1] Surgical resection in the thoracic cavity is often complicated by the tumor's proximity to or involvement with the great vessels, cardiac structures, esophagus, airway, and critical nerves. These operations

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Abbreviations and Acronyms
CPB = cardiopulmonary bypass
CT = computed tomography
PPB = pleuropulmonary blastoma
SEF = sclerosing epithelioid fibrosarcoma
ToF = tetralogy of Fallot

are further complicated by cardiac and respiratory motion.[2] Iatrogenic injury is rare but can be catastrophic.[2] If an injury occurs, prompt and effective control of bleeding and repair of the injury are paramount to avoid undue morbidity and mortality.[2] Mobilizing thoracic structures to gain optimal anatomic exposure can make complementary procedures that entail complex resections with aberrant anatomy feasible.

Multidisciplinary surgical teams are common in other complex pediatric operations; examples include operations for tracheoesophageal fistula/esophageal atresia, congenital diaphragmatic hernia, congenital airway malformations, cloaca repairs, and the separation of conjoined twins. Diverse teams in surgical oncology facilitate radical or en bloc resections and allow synchronous reconstruction[3]; for example, vascular surgeons may assist with resection and immediate reconstruction of the hepatic artery and portal vein during pancreatic adenocarcinoma resections.[4] In oncologic cases where a tumor or thrombus extends above the diaphragm, cardiac surgery involvement is often recommended to facilitate median sternotomy, vessel cannulation, and the use of cardiopulmonary bypass (CPB).[3] Cases involving intracardiac neoplasms or thrombus typically require CPB and may require extensive vascular manipulation.[3] Congenital cardiac surgeons have familiarity with thoracic and vascular structures in both adult and pediatric patients. Because of this experience, cardiac surgery involvement in thoracic surgical oncology is beneficial.

Here we present 4 cases that either required or benefited from cardiac surgery collaboration. The families of all 4 patients included in this study were contacted via phone and gave their verbal consent for inclusion in this report; Institutional Review Board approval was not required.

CASE 1

A 17-year-old male underwent resection of a 16 × 15.5 × 11 cm right chest synovial sarcoma (Figure 1, *A*) following neoadjuvant chemoradiation. A computed tomography (CT)-guided biopsy of his thoracic mass established the diagnosis of synovial cell sarcoma. This biopsy was performed 6 months prior to surgical resection and was the basis for the neoadjuvant therapy provided. He underwent 16 weeks of neoadjuvant ifosfamide and doxorubicin treatment as well as proton radiation therapy (5000 cGy in 25 fractions). Imaging revealed atelectasis with a leftward shift of the aorta, superior vena cava, right-sided pulmonary veins, and trachea. Also noted were encasement of brachiocephalic, subclavian, and right internal mammillary arteries, as well as encasement of the phrenic nerve and vagus nerve and possible brachial plexus involvement (Figure 1, *B*). Resection was performed by a multidisciplinary team including cardiothoracic, vascular, neuroplastics, and pediatric surgery. The surgical approach via a hemi-clamshell approach allowed for complete R0 resection including right upper lobectomy, pleurectomy, phrenic and vagus nerve resection, and partial pericardiectomy. The right subclavian artery was reconstructed using an 8-mm ringed synthetic graft. The patient was extubated in the operating room, had an uneventful hospital course, and was discharged on postoperative day 10. He received an additional month of adjuvant chemotherapy after discharge.

The patient did not experience local recurrence but underwent an additional resection at 25 months following the index operation due to a distant pleural nodule that was identified as a synovial sarcoma on biopsy. The re-resection consisted of a right posterolateral thoracotomy, radical total visceral and total parietal pleurectomy and decortication, diaphragm reconstruction with porcine dermal graft, and thoracic duct ligation. He recovered from this surgery uneventfully and was discharged on postoperative day 5. At 23 months postoperatively, a distant pleural nodule was identified 2 years after diagnosis in the setting of an initial pleural effusion.

CASE 2

A 14-year-old female had a primary resection of a 17.6 × 10.5 × 8.1 cm sclerosing epithelioid fibrosarcoma (SEF) of the posterior mediastinum (Figure 2, *C*). She underwent a CT-guided biopsy of her thoracic mass 1 month prior to surgical resection, which established the diagnosis of SEF. Preoperative radiologic analysis showed a calcified, posterior mediastinal mass originating from the vertebral body. The mass compressed the aortic arch, right and left pulmonary arteries, trachea, and esophagus with lateral displacement (Figure 2, *A* and *B*). The patient underwent a left posterolateral thoracotomy in the fourth interspace initially, but because of the large and densely adherent nature of the mass, the third interspace was ultimately accessed as well. Surgeons preserved all thoracic anatomy except a small aortic collateral vessel at the level of T3 despite extensive periaortic and posterior mediastinal dissection. Surgical margins were positive at the posterior aspect of the tumor where it directly abutted the anterior vertebral column.

The patient was extubated intraoperatively, had an uneventful postoperative course, and was discharged on hospital day 5. She was treated with adjuvant proton radiotherapy (4500 cGy with a focal boost of 5580 cGy in 31 fractions)

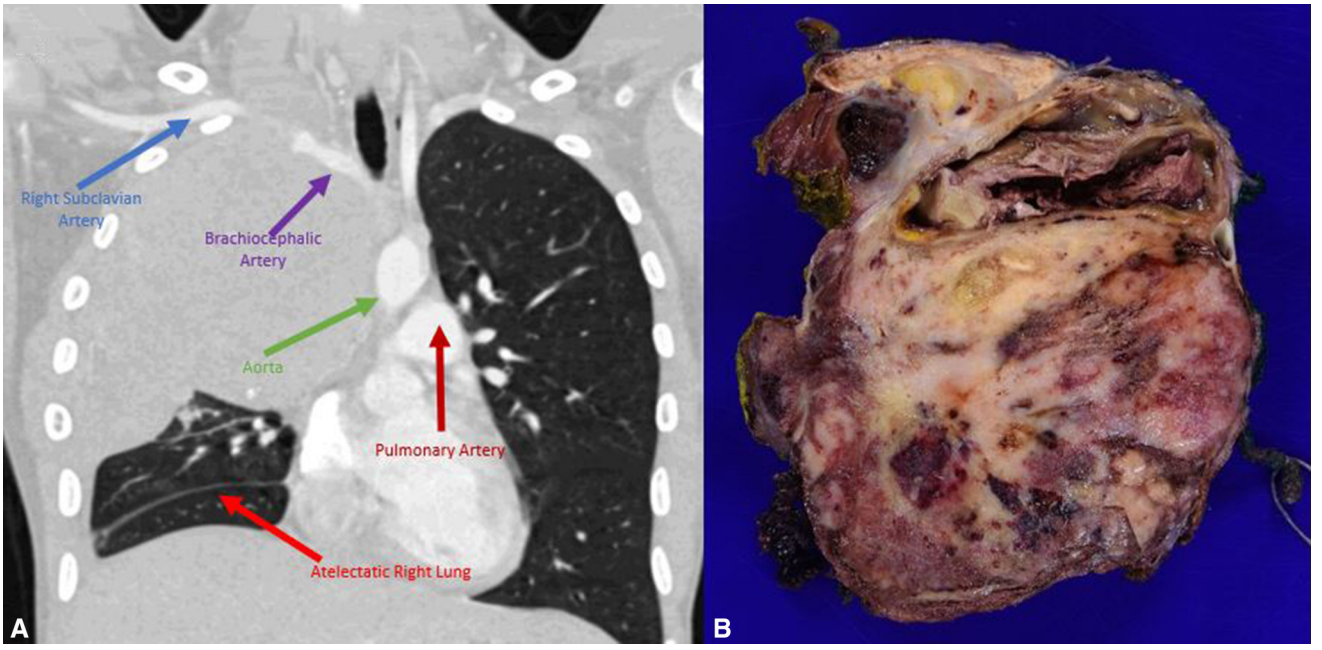

FIGURE 1. A, Large right-sided partially calcified mass causing atelectasis and leftward deviation of the aorta, superior vena cava, right-sided pulmonary veins, and trachea, with encasement of the brachiocephalic, subclavian, and right internal mammillary arteries. B, Gross tumor showing an encapsulated and partially hemorrhagic nonviable mass with portions of the right vagus, intercostal, and phrenic nerves.

and had no clinical or radiologic evidence of recurrence at 1 year postoperation.

CASE 3

An 18-month-old female underwent synchronous removal of an incidentally detected 11.5 × 9 × 5.5 cm type IR pleuropulmonary blastoma (PPB) (Figure 3, *B*) at the time of tetralogy of Fallot (ToF) repair. The patient immigrated the United States and was symptomatic and cyanotic when care was established. Oxygen saturation in her lower extremities was 70%, and she had a large right-to-left shunt detected on echocardiography. Preoperative radiologic analysis revealed a left-sided cystic lesion causing atelectasis and a rightward mediastinal shift (Figure 3, *A*). Differential diagnoses of the lesion included pneumocele, pneumothorax, and neoplasm. Because the patient was hemodynamically stable, thoracostomy tube was avoided, with a plan for exploration at the time of surgical repair of the cardiac defect. To avoid the risk of creating a pneumothorax or bronchopleural fistula, preoperative instrumentation was avoided. ToF repair was performed via median sternotomy with a polyester patch (Dacron; Maquet Cardiovascular), closure of the ventricular septal defect, and a transannular patch of pulmonary homograft

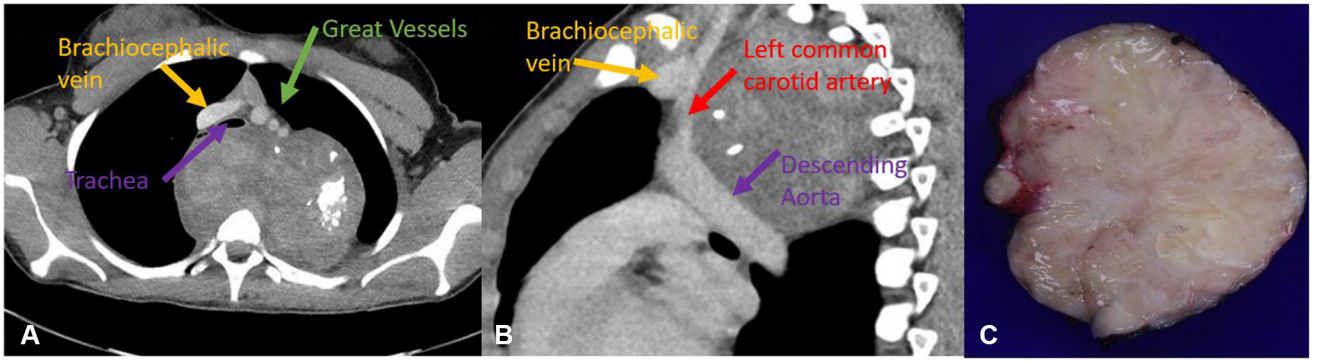

FIGURE 2. A and B, Partially calcified posterior mediastinal mass that is mostly left of the midline abutting the upper thoracic vertebral bodies and left pleura, with anterior displacement of the trachea, great vessels, and esophagus. C, Gross specimen showing an encapsulated, multinodular mass with hybrid features of sclerosing epithelioid fibrosarcoma and low-grade fibromyxoid sarcoma.

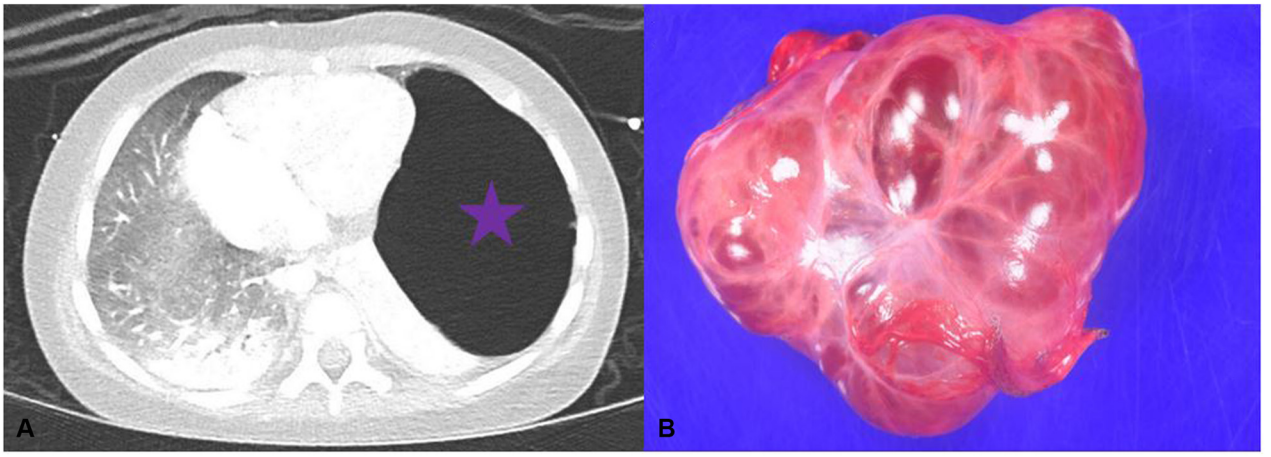

FIGURE 3. A, Cystic mass in the left thoracic cavity causing compressive atelectasis of the left upper and lower lung lobes. Moderate right-sided mediastinal shift is also seen (*purple star* is located int he center of the cyctic mass). B, Gross specimen showing a uniloculated cystic mass with 3 staple lines present.

to augment the right ventricular outflow tract. Echocardiography confirmed the quality of ToF repair. Next, anticoagulation was reversed with protamine administration, and attention was directed to the left hemithorax. The left pleural space was opened, and the mass was inspected and found to originate from the lingula. It was excised using endoscopic staplers, with limited resection of lung parenchyma. Following excision, the left lung was ventilated and appeared to have normal function with no air leak. Both mediastinal and pleural drainage tubes were placed.

The patient was extubated in the operating room and had an uneventful hospital course. She was discharged on hospital day 10. Pathology revealed a type IR PPB with negative margins. Postoperative X-ray on the day of surgery showed reinflation of the left lung with no evidence of pneumothorax or effusion. Follow-up chest CT scan showed no evidence of recurrence at 1 year postoperatively.

CASE 4

This 11-year-old female underwent resection of a 15 × 19 × 22 cm Wilms tumor with focal anaplasia that extended into the inferior vena cava (IVC) (Figure 4, *B*). An interventional radiography–guided biopsy of her perirenal mass established the diagnosis of a stage 4R Wilms

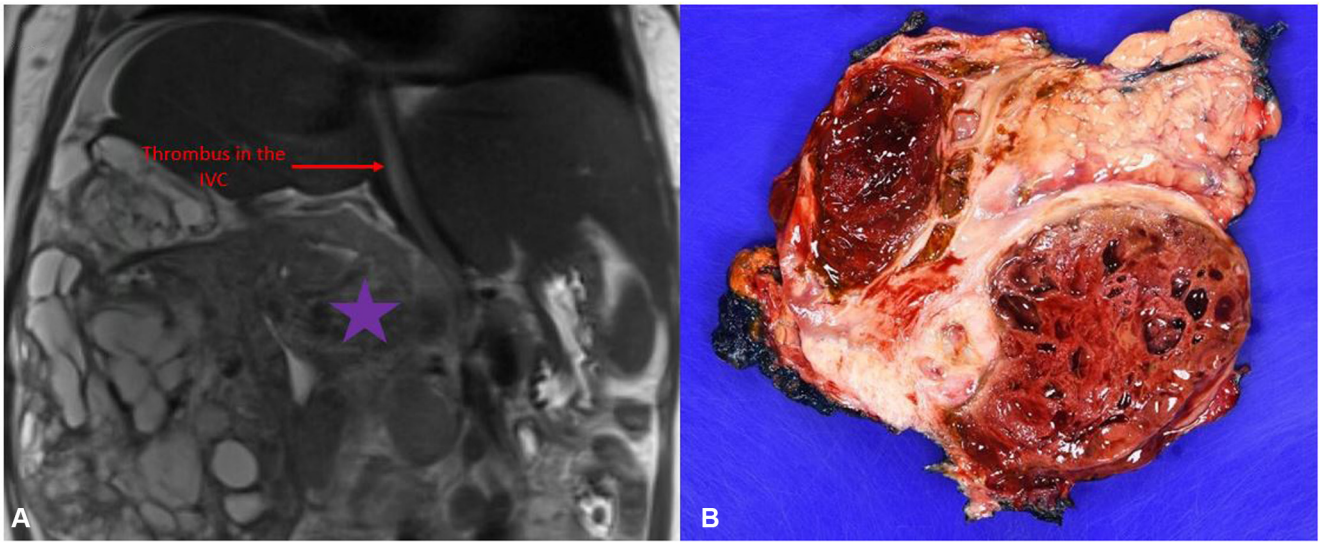

FIGURE 4. A, Computed tomography scan showing a heterogeneous mass partially displacing the liver and pancreas replacing right kidney with tumor extension into the right renal artery and vein, right adrenal gland, and psoas muscle. A thrombus is visualized in the intrahepatic inferior vena cava (*purple star* marks the location of the Wilms tumor). B, Gross specimen of Wilms tumor with residual positions of the liver and kidney and tumor extension into the ureter.

tumor. This biopsy was performed 2 months prior to surgical resection, and she was treated with 10 weeks of neoadjuvant doxorubicin, vincristine, and dactinomycin. Preoperative imaging showed a heterogenous mass arising in the right kidney with invasion into the right adrenal gland, right psoas muscle, and right hepatic lobe. Near occlusion of the IVC also was present from a tumor thrombus that extended into the right atrium (Figure 4, *A*). Intraoperative transesophageal echocardiography showed a thrombus traversing from the right renal vein to the level of the eustachian valve. To obtain a complete resection, the ureter, gonadal vein, adrenal gland, and renal artery were sacrificed. In addition, eradication of the posterior aspect of the tumor necessitated resection of a 5 cm × 5 cm portion of the diaphragm and a portion of the right lobe liver. Rummel tourniquets were used to occlude the distal right renal vein and proximal suprahepatic IVC to remove the nonviable tumor and infiltrated portion of the vessel with CPB standby. Renal vein repair was performed primarily, and the diaphragm was patched using 1-mm-thick synthetic material. On pathologic analysis, the superior, lateral, and medial perinephric tissue margins as well as the renal vein margin were positive for nonviable tumor.

The patient was extubated intraoperatively, had an uneventful postoperative course, and was discharged on postoperative day 5. She was treated with 10 weeks of adjuvant vincristine, dactinomycin, doxorubicin, cyclophosphamide, and etoposide. She also received 1050 cGy in 7 fractions to the whole abdomen with a focal cone down boost of 900 cGy in 5 fractions (total 1950 cGy in 12 fractions). Follow-up exam revealed no clinical or radiologic evidence of recurrence at 6 months postoperation.

DISCUSSION

Rare and invasive tumors are challenging to manage and often require a multidisciplinary approach for adequate local control. Here we present examples of several different tumor types, each of which required a multidisciplinary surgical team to achieve the best possible results.

Synovial sarcoma accounts for 14.7% of intrathoracic sarcomas, making it the most common subtype.[5] Synovial sarcoma is an aggressive tumor, notorious for high rates of local recurrence and metastatic spread, and is associated with high rates of mortality.[5] For localized tumors, negative margins are paramount for disease-free survival.[5] A retrospective analysis of 13 patients with this pathology displayed 2-year and 5-year survival rates of 58.3% and 30%, respectively. In this cohort, 11 patients (84.6%) experienced recurrence, 7 of whom with intrathoracic recurrence or pulmonary metastasis.[5] The authors stressed the importance of a multidisciplinary collaboration among surgeons, radiologic oncologists, oncologists, and other medical physicians to effectively treat these patients.[5] An additional study of 20 patients with thoracic synovial sarcoma showed a 5-year survival of 22%, with 75% of the cohort experiencing recurrence.[6] The sole significant predictor of disease recurrence was incomplete resection of the primary tumor.[6] Therefore, complete resection is compulsory for disease-free survival.

In our case 1, we achieved a complete resection despite the patient's extensive tumor burden. This would have been impossible without the careful logistics and collaboration among surgeons. Aggressive resection conceivably improved the prognosis for this patient and allowed him to have a good quality of life following surgery. Although this patient experienced recurrence 2 years after surgery, he underwent metastectomy via right thoracotomy, and another disease-free interval is hoped for.

In case 2, the patient was diagnosed with SEF. Only slightly more than 100 cases of SEF were reported between 1995 and 2021.[7] Most cases occur in adults, with a propensity for the lower extremities.[7] Owing to the rarity of this pathology, the optimal treatment has not yet been clearly defined. SEF is not chemotherapy-responsive, and recurrence rates are high, even in patients treated with aggressive multimodal therapy.[7] A study of 13 histologically confirmed cases of SEF included 7 patients with lower limb disease, 5 patients with thoracic disease, and 1 patient with disease in the neck.[7] Of these cases, 11 patients presented with locally contained disease and underwent radical resection. All patients experienced local recurrence, and 10 patients developed metastatic disease at a median of 12.4 months following their initial diagnosis. Of note, all 11 patients had microscopically positive margins at time of resection,[7] confirming the tendency for SEF to recur and metastasize. In 1 reported case with right pulmonary artery encasement and superior vena cava, aorta, and left bronchus impingement, the patient achieved 69-month disease-free survival.[8] Of note, this resection involved repair of the right pulmonary artery and superior vena cava, and thus cardiothoracic surgeons were involved and CPB was used.[8] Our case required extensive posterior mediastinal dissection near branches of the aorta. The safe conduct of this operation, aided by cardiac surgery, allowed safe and aggressive resection with CPB standby and the capacity for repair of the aorta.

In case 3, the patient underwent synchronous ToF repair and PPB resection. PPB is a rare and potentially malignant tumor of the lung or pleura occurring in young children.[9] Despite its rarity, it also is the most common primary pulmonary malignancy in children.[9] PPB is believed to begin as type I and may progress or regress (type Ir). Type Ir has no malignant potential and is not associated with mortality.[9] Alternatively, PPB can progress from type I to type II (with an expanded solid component) or type III (entirely solid), becoming an aggressive sarcoma

necessitating multimodal therapy and carrying a poor prognosis owing to the possibility of local recurrence or metastatic spread.[9] Achieving optimal resection for type II tumors, type III tumors, and tumors larger than 10 cm is difficult[9]; therefore, we elected to resect the PPB at the time of ToF repair. Surgical treatment is the standard of care for type I PPB and often consists of a large wedge resection, frequently using an open approach.[9] This was the approach used in our case. However, owing to the synchronous presentation with symptomatic ToF, resection via median sternotomy was undertaken. It was unclear whether the patient's hypoxemia was due to underlying ToF, left lung compression from the PPB, or both.

Advantages of synchronous cardiac surgery and lung resection can be extrapolated from the adult literature.[10] In 10 patients undergoing synchronous anatomic lung resection and aortic valve replacement and/or coronary artery bypass grafting, there were no deaths or reoperations for bleeding.[10] This synchronous approach spared our patient from an additional operation, as well as the toxicities of the chemotherapy agents and radiation treatment she would have required had the PPB progressed to type II or III. In addition, the patient's recovery from ToF might have been aided by relieving the local compressive effects or impeded ventilation caused by the PPB. An alternative strategy might have been to sample the lesion at the time of the operation and leave the bulk of the tumor for resection after definitive pathologic results were available. This approach likely would have slowed the patient's recovery following ToF repair and confounded issues surrounding hypoxemia after ToF repair, such as right ventricular noncompliance and shunting at the atrial level. In this case, leaving the tumor would have delayed the PPB surgery and necessitated an additional operation that could be made more difficult by the existence of adhesions. This series of operations also would require an additional anesthetic induction, and the end result would be congruent to the single operation performed by the cardiac surgeon in this case. This child received definitive treatment through her preexisting median sternotomy, and because her lesion was type IR PPB, she is free of lung pathology.

In case 4, the patient was diagnosed with a Wilms tumor that had tumor-containing thrombus traversing the IVC into the right atrium. In such cases when a Wilms tumor and accompanying thrombus are fully resected, prognosis then align with prognoses in children with standard tumors.[11] In a study of 24 patients with Wilms tumor accompanied by tumor thrombus extending into the IVC, 9 required CBP intraoperatively.[11] In another study that analyzed 40 surgical patients with Wilms tumor, primitive neuroectodermal tumor of the kidney, hepatoblastoma, or adrenocortical carcinoma with tumor or tumor-related thrombus extending into the inferior vena cava, 6 patients (15%) underwent CBP. The authors advocated for cardiac surgery involvement and CPB in all cases with supradiaphragmatic thrombus.[3] In our series, selective snaring of the suprahepatic IVC and renal vein allowed us to avoid CPB. The diseased vein was resected and repaired primarily by the abdominal pediatric oncologic surgeon. This vascular management facilitated a more complete resection in this very locally advanced case.

When cancers arise within or extend into the thoracic cavity, cardiac surgeons may join the multidisciplinary team. A review of 22 patients with intrathoracic paragangliomas showed that 9 required CPB.[12] Of note, of the 13 patients who did not require CPB, 9 had a tumor associated with the great vessels, 2 had pericardial involvement, and 1 had a tumor associated with the right atrial groove. The discerning use of CPB in these complex dissections exemplifies the technical skills and comfortability that cardiac surgeons bring to multidisciplinary teams.

CONCLUSIONS

Pediatric patients may present with locally advanced heterogenous neoplasms. Thorough familiarity with the anatomy of the mediastinum, thoracic hilum, and great vessels can facilitate safe and complete resection in all cases. The creation of a multidisciplinary team ensured that optimal resections were performed in these complex patients. These cases demonstrate the value of cardiothoracic surgery consultation in the management of complex oncologic tumor resection of the chest.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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Key Words: congenital, thoracic, education, vascular, oncology