Complex Head and Neck Resection, Reconstruction, and Rehabilitation in Children

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KEYWORDS

• Pediatric • Head and neck • Tumor • Cancer • Ablation • Reconstruction
• Rehabilitation

KEY POINTS

- Pediatric head and neck tumors are uniquely challenging because of their complex 3-dimensional anatomy and the need to balance oncologic goals with functional morbidity.
- Multidisciplinary preoperative consultations include speech and language pathology for voice, swallowing, tracheostomy education, hearing, dental, and psychosocial assessments, in addition to ablative and reconstructive surgical teams.
- Intraoperative surgical planning considers airway interventions including short-term tracheostomy use as well as ventilating ear tube placement and nasolacrimal duct stenting.
- Reconstruction in pediatric patients should consider virtual surgical planning, future growth, nerve reconstruction, and dental restoration options.
- Free flap reconstruction can be used safely and effectively in children to restore craniofacial form and function.

INTRODUCTION

Pediatric head and neck tumors are uncommon but the consequences of radical resection are extensive. These tumors, benign and malignant, are uniquely challenging because of their proximity to critical functional and neurovascular structures and intimately affect speech, swallowing, voice, breathing, hearing, and vision. In addition, the psychosocial and emotional trauma from the cosmetic and functional consequences can be enduring. Their relative rarity limits surgeon experience and requires a focused effort to develop individual and programmatic expertise. A practiced

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multidisciplinary team can facilitate smooth preoperative evaluations, efficient coordinated operative procedures, comprehensive rehabilitation, and recovery, as well as optimal oncologic outcomes.

Primary considerations

There is an extensive range of tumors, benign and malignant, odontogenic and nonodontogenic, which cause swelling of the head and neck in children and young adults (**Table 1**). The treatments range from observation to medical management, curettage, and radical resection. The morbidity of aggressive treatment in children must be balanced with the risk of recurrence over the lifespan of our young patients. Though this list is far from complete, there are some pathologies that deserve special consideration.

Desmoid Tumors

Desmoid tumors are rare, benign, locally aggressive, and infiltrative tumors. They typically occur in children younger than 11 years and equally in male and female patients. Tumor genetics may reveal sporadic desmoid tumor with β-catenin mutation (*CTNNB1*) or familial adenomatous polyposis (FAP)-associated desmoids with a mutation in chromosome 5 associated with the APC gene.[1] Desmoid tumors of the head and neck typically appear as a painless mass adjacent to the mandible or cervical region but may also be found in the skull base or parapharyngeal regions. The diagnosis is challenging and either open biopsy or core needle biopsy is recommended. Open biopsy is preferred to perform tumor mutational analyses.[2] Historically, surgical treatment has been preferred with negative microscopic margins. However, this treatment plan is particularly difficult in the head and neck because of the extreme risk of cosmetic and/or functional deficits either from local tumor infiltration of critical head and neck structures or iatrogenic from aggressive surgical resection. More recent studies suggest that resection margins do not clearly correlate to prognosis and recurrence rates after surgical resection are high (20–70%).[3] Currently, the global Desmoid Tumor Working Group recommends an initial period of active surveillance with close-interval imaging and if aggressive tumor progression occurs, escalating treatment to medical treatment (conventional chemotherapy, antihormonal therapies, nonsteroidal anti-inflammatory drugs, or tyrosine kinase inhibitors) or surgical resection or radiation therapy.[2] Overall, there is a lack of evidence for a specific treatment algorithm and the current trend is shifting to a more conservative approach, with active surveillance and medical therapies when indicated.

Osteosarcoma, ewing sarcoma, and chondrosarcoma

Sarcomas are the most common primary bone tumor in children but rarely affect the head and neck. Owing to the low incidence, treatment paradigms often group these tumors with other pediatric head and neck malignancies or other subsite sarcomas. The incidence of bony sarcomas is increased in children with tumor predisposition syndromes or a history of head and neck radiation. Aggressive primary surgical resection with wide margins is preferred for maximum survival benefit but this is difficult in the pediatric head and neck because of the proximity of critical neurovascular structures, smaller pediatric anatomy, and cosmetic and functional morbidities.[4] Multimodality therapy is used in the treatment of bony sarcomas, but disease-specific survival is highest with surgery alone, reflecting the survival benefit of surgically resectable tumors.[5]

Rhabdomyosarcoma

Rhabdomyosarcoma is classified as a soft tissue sarcoma and is one of the most common tumors of childhood. The head and neck is a common site and tumors are

Table 1
Differential diagnosis of pediatric mandible and maxillary tumors

Benign	Malignant	Dental Tumors
Juvenile ossifying fibroma	Osteosarcoma[a]	Ameloblastoma
Osteoma	Chondrosarcoma[a]	Dentigerous cyst
Osteoblastoma	Malignant ameloblastoma	Keratocystic odotogenic tumor
Desmoid fibromatosis	Ewing sarcoma[a]	Adenomatoid odontogenic tumor
Desmoid tumor	Squamous cell carcinoma[a]	Calcifying epithelial odontogenic tumor
Desmoplastic fibroma	Rhabdomyosarcoma (invasion)[a]	Squamous odontogenic tumor
Fibromyxoma	Metastases[a]	Odontoma
Central Giant Cell Granuloma		Ameloblastic fibroma
Fibrous dysplasia		Ameloblastic fibrosarcoma[a]
Teratoma		Odontogenic myxoma
Lymphangioma		Cementoblastoma
Vascular tumor		Odontogenic fibroma
		Odontogenic fibromyxoma

[a] Malignancy.

bimodal in incidence, occurring most frequently in children younger than 5 years and adolescents aged 10 to 18 years.[6] Rhabdomyosarcoma is divided into the following categories: (1) embryonal, (2) alveolar, and (3) pleomorphic. This categorization is crucial for prognosis, with alveolar-type carrying the worst prognosis. Beyond histology, molecular diagnostics assessing for common tumor mutations also carry significant impact on prognosis, with the FOX-01 driver mutation leading to decreased survival and shortened disease-free intervals.[7] Treatment is multimodal and tumors are grouped based on resectability: group I is completely resected with clear microscopic margins, group II is gross tumor resection with residual microscopic disease, group III is gross residual disease or biopsy only, and group IV is distant metastases at presentation. Aggressive surgery is used in the head and neck for salvage when there is residual disease to reduce the radiation dose and long-term side effects, but often radiation therapy is necessary for group II patients or higher.[8]

Odontogenic Tumors

Many destructive masses of the mandible or maxilla are of odontogenic origin. Although MRI may help delineate the solid or cystic nature of the mass, a computed tomography scan is often equivocal or misleading. Cystic odontogenic cysts may be benign, such as odontogenic keratocyst, eruption cyst, and unicystic ameloblastoma. Odontogenic malignancies include malignant ameloblastoma, ameloblastic fibroma, and odontogenic myxoma. Diagnosis typically requires open biopsy and treatment may range from observation to enucleation and curettage, to segmental resection.[9,10] Multidisciplinary care is necessary with experienced pathology, oral surgery, otolaryngology, and reconstructive surgery to ensure the care plan is neither overly conservative nor too aggressive.

PREOPERATIVE PLANNING

Multidisciplinary care relies on consistent and complete communication between services. Consultations with ablative and reconstruction teams include head and neck surgery, craniofacial and plastic surgery, oncology, neurosurgery, ophthalmology, dental surgery, anesthesia, speech and language pathology, child life, and social work. Clinical nursing and surgical coordinators are invaluable patient contacts to help coordinate visits when possible because of the extensive time burden of these consultations and associated imaging studies.

Surgical timing is very difficult in complex cases and is multifactorial. Early discussion between ablative and reconstruction teams initiates virtual surgical planning (VSP) when indicated, as these implants typically require 1-2 weeks for design and fabrication. In addition, many oncology patients require local control with surgical resection at a specific interval following induction chemotherapy treatments. Finally, coordinating multiple surgeons and surgical teams is always challenging. For this reason, we recommend a consistent team that is practiced and well-coordinated.

We employ a short preoperative planning session the week before the procedure with the entire intraoperative team: anesthesiology, nursing and scrub technician teams, and surgeons. The COVID-19 pandemic prompted us to switch to virtual meetings, which has allowed more regular and complete attendance. We find these meetings to be feasible and highly effective in ensuring a well-coordinated case. The team agrees and publishes a written plan that confirms the sequence of procedures, room setup, possibility of simultaneous tumor extirpation and free flap harvest, availability of all instruments and implants, specimen handoff with anticipation of frozen section analyses, perioperative disposition, and the need for staged procedures or adjuvant therapy. **Table 2** and **Fig. 1** are examples from the preoperative plan for surgical case 1.

SURGICAL CASES

Case 1: Maxilla

A 13-month-old infant boy was evaluated for a rapidly enlarging right facial mass. He underwent evaluation by oncology and otolaryngology and decision was made to proceed with sublabial biopsy which confirmed desmoid fibromatosis. Because of patient age and cosmetic risk of radical resection, decision was made for short-interval observation. Within 3 weeks, the tumor was noted to enlarge, and medical therapy with doxorubicin was initiated. The tumor improved minimally with 3 cycles of doxorubicin and decision was made to proceed with radical resection.

The patient underwent tracheostomy, right total maxillectomy, myringotomy and ventilation tube placement, right nasolacrimal duct stenting, and anterolateral thigh free tissue transfer. He tolerated the procedure well and was transferred to the intensive care unit for flap monitoring and fresh tracheostomy precautions. He did well postoperatively and was decannulated on postoperative day (POD) 15 and discharged home on day 16 with a soft diet. He is doing very well at 9 months after treatment with no evidence of recurrence (**Fig. 2**).

Case 2: Mandible

An 11-year-old girl was referred by the dental service for a rapidly enlarging right mandible tumor. Dental biopsy demonstrated ameloblastoma and pathology was confirmed with in-house pathologist. Decision was made to proceed with segmental resection with VSP. A temporary tracheostomy was placed at the time of the procedure for the perioperative period. Custom cutting guides and reconstruction plates were

Table 2
Example of preoperative planning session

Team	Procedure	Instruments and Equipment
Anesthesiology	Intubate, basic IV access	
Head and neck	Tracheostomy, myringotomy, and ventilating tube	5.0 Cuffed Tracheostomy
Anesthesia and nursing	Turn bed, padding/positioning, central lines, arterial line, and foley	U/S Probe cover
Head and neck Plastic Reconstruction	Tumor resection RIGHT lower ext. free flap preparation	Simultaneous: 2 tables, 2 set ups
	Marginal frozen section, timeout before reconstruction	
Dental, oculoplastics	Dental implant to fibula and/or nasolacrimal duct stenting	
Pathology	Frozen section confirmed	Clean set up
Plastic Reconstruction/ Craniofacial teams	Free flap inset Feeding tube insertion	
Anesthesia and nursing	Transfer to ICU	

designed and used for an osseous-only fibula free flap, which was harvested simultaneous to tumor resection. Dental implants were placed in the fibula before flap transfer and anastomosis to the facial vessels and flap inset. Also, nerve interposition graft was used to reinnervate the proximal inferior alveolar nerve and distal mental nerve segments.

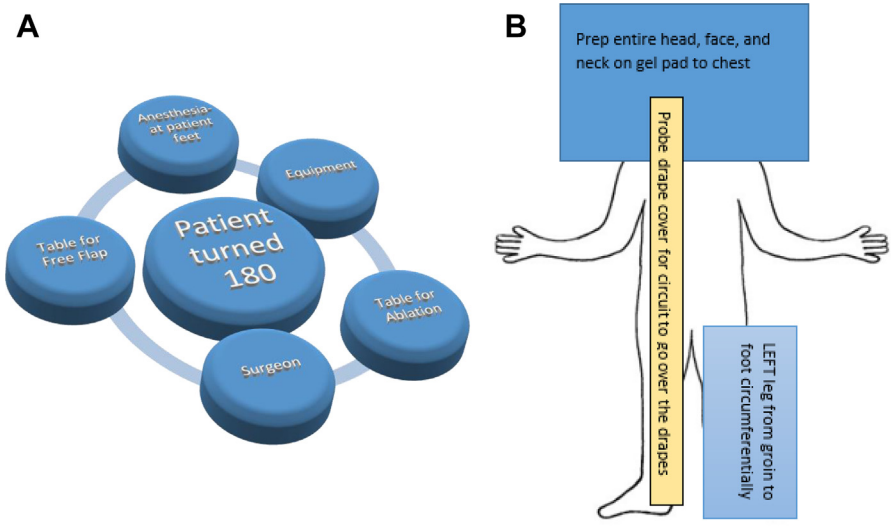

Table & Personnel arrangement Draping plan

Fig. 1. Example of preoperative planning session figures. (*A*) Table and Personnel arrangement. (*B*) Draping plan.

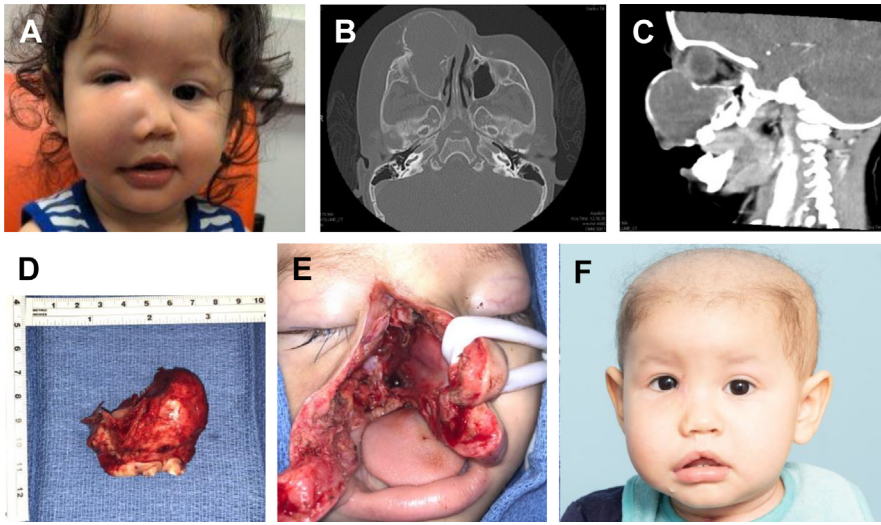

Fig. 2. Maxilla. (*A*) Preoperative photo. (*B*) Axial bone CT scan with large right maxillary tumor. (*C*) Sagittal soft tissue CT scan with large right maxillary tumor exerting inferior pressure on the globe. (*D*) Tumor extirpation. (*E*) Intraoperative surgical defect. (*F*) One-month postoperative photo.

The patient remained in the intensive care unit for recovery and fresh tracheostomy protocol. She was decannulated on POD 9 and discharged home on POD 10. She was tolerating full liquid oral diet at the time of discharge (Fig. 3).

Case 3: Neck/Skull Base, Lower Cranial Nerves

A 17-year-old male with a known history of type 1 neurofibromatosis (NF1) presented with a painful, enlarging left neck mass, dysphagia, and voice change. A PET scan showed FDG-avidity. Ultrasound-guided fine needle aspiration demonstrated concerning features for malignant peripheral nerve sheath tumor including high cellularity and loss of S100 IHC expression. He had normal vocal fold mobility on preoperative in-office laryngeal stroboscopy. In anticipation of voice and swallowing compromise following resection, the patient underwent radical neck dissection with transcervical radical tumor resection and sacrifice of the vagus nerve from the clavicle to skull base. Simultaneous recurrent laryngeal nerve (RLN) reinnervation was performed with direct anastomosis of the ipsilateral ansa cervicalis to the RLN, and intraoperative injection of the left vocal fold using carboxymethylcellulose. Final pathology revealed atypical neurofibroma with all lymph nodes demonstrating benign reactive follicular hyperplasia.

He was admitted to the pediatric intensive care unit and transferred to the floor on POD 3. Postoperative videofluoroscopic swallow study showed tracheal aspiration of thin and nectar-thick liquid barium with abundant residual in the piriform sinuses. A gastrostomy tube (G-tube) was placed on POD 17 and he was discharged home on POD 19. He continued to have aspiration of secretions and poor oral feeding. At 5 months postoperatively, he underwent in-office transcervical hyaluronic acid injection. At 6 months, he reported normal voice and was able to tolerate full oral diet without coughing (Fig. 4).

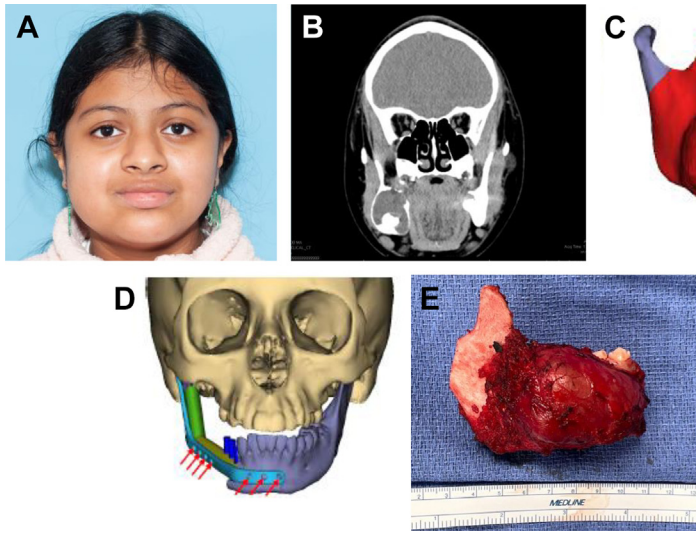

Fig. 3. Mandible. (*A*) Preoperative photo. (*B*) Soft tissue coronal CT scan demonstrating large cystic mass in right hemi mandible. (*C*) Virtual surgical planning images showing planned resection (*red*). (*D*) Virtual surgical planning with custom plate (*teal*), fibula segments (*yellow and green*), and dental implants (*blue posts*). (*E*) Tumor extirpation after segmental mandibulectomy, lateral view, with clear margins on frozen examination.

POSTOPERATIVE CONSIDERATIONS

Careful postoperative management is critical to successful reconstruction of the pediatric head and neck tumor patient. Patients are admitted to the acute care or critical care units with adequate analgesia to minimize risk to the reconstruction. Pediatric patients are often less likely to be compliant with immediate postoperative restrictions. Sedation is used for 24-72 hours if needed in the setting of a fresh free flap anastomosis and fresh tracheostomy. Short-term tracheostomy decannulation protocol may be used to rapidly wean airway support. Daily transtracheal pressure monitoring can be used to guide tracheostomy downsizing and capping trials to ensure safe and timely decannulation before discharge.[11] Experienced pediatric speech and language pathologists are critical for guiding decannulation as well as advancing swallow evaluations and oral feeding and in a concurrent fashion. This optimizes the patient's success for decannulation and full oral diet at the time of discharge. Patients typically spend 5-7 days in the critical care unit and another 5-7 days in the acute care setting.

Other pearls for the care of the pediatric patient are incorporating child life specialists and dedicated occupational and physical therapists in the immediate postoperative period. Children rehabilitate quickly when the patient and caregivers are actively engaged in their recovery. Patients rarely need sustained physical and occupational therapy in the outpatient setting after discharge.

LONG-TERM SURGICAL OUTCOMES AND REHABILITATION

Head and neck tumors in pediatric patients are particularly challenging because of the pathologic variability and rarity of any single pathology. In addition, reconstruction of surgical defects in the growing craniofacial skeleton and the restoration of form and function is more difficult in smaller patients with fewer donor site options, smaller

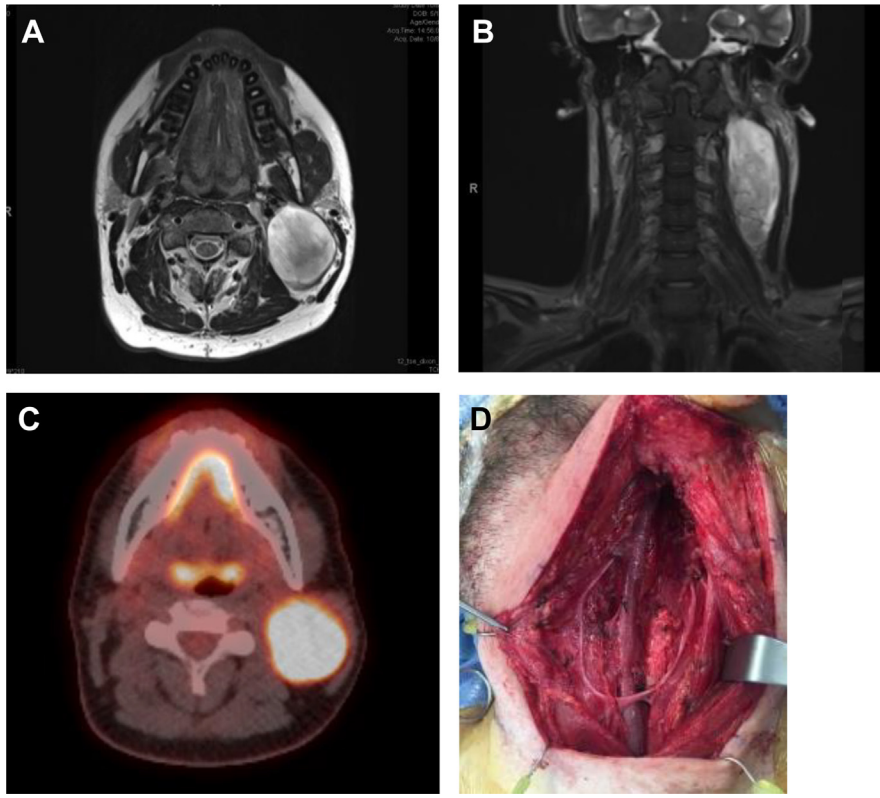

Fig. 4. Neck/skull base, lower cranial nerves. (*A*) Axial T2 weighted MRI with large left neck mass. (*B*) Coronal T2 weighted MRI demonstrating left neck mass extending along carotid space to skull base. (*C*) FDG PET with PET-avid lesion in left neck. (*D*) Intraoperative photo of left ansa-cervicalis post-tumor resection before RLN-ansa anastomoses.

anastomoses, and unpredictable postoperative compliance.[12,13] On the other hand, many pediatric head and neck tumors are benign, and our patients are less likely to have received preoperative chemotherapy and radiation therapy than adult head and neck patients. Overall, there are excellent options and outcomes for reconstruction including free tissue transfers in pediatrics for maxillary, mandibular, and other craniofacial defects.

Free flap reconstruction can be used safely and effectively for pediatric maxillary and mandibular defects with few complications.[10,12,14–19] In one multi-institutional study of pediatric free fibula reconstruction, minor short-term and long-term complications included minor wound dehiscence (7.8%) and cellulitis (4.5%) and lower incidence of major complications of wound infection (6.7%), dehiscence (2.2%), or flap failure (1.1%). Longer-term severe speech deficit was uncommon (6.5%). There were few complications to the donor site as well and no patients required orthopedic intervention as gait and limb length were normal. When compared to adult patients, there were fewer short-term and long-term complications[14] Another study reported an overall success rate of 95.6% for free tissue reconstruction of pediatric head and neck defects.[19] Complications were significantly increased in patients aged 5-9 years, which may be attributed to the smaller vessel size, but this conclusion is limited by the very rare incidence of flap failure.[19]

A primary concern for pediatric mandible and maxilla reconstruction is disruption of the growth of the craniofacial skeleton. Several studies have reported that most free flaps continue to grow in the recipient area.[19–22] Neo-growth of free fibula mandibular reconstruction is greatest when the condyle is preserved, mandible epiphyseal growth plate is preserved, and a younger age, hence greater growth potential of the donor fibula.[20] The growth of the mandibular reconstruction impacts long-term craniofacial development and mandibular symmetry, which is also significantly impacted by dental restoration. Proper dental occlusion encourages symmetric mandibular growth and when done concurrently, patients with dental rehabilitation at the time of free fibula reconstruction do not have long-term craniofacial abnormalities[14] Unfortunately, dental restoration is expensive and financial barriers lead most patients to undergo delayed dental implants.

Additional considerations should be given to peripheral sensory nerve restoration to improve quality of life after radical mandible resection. There is a growing trend and convincing evidence that immediate inferior alveolar nerve allograft reconstruction results in complete functional sensory recovery in pediatric patients.[23]

SUMMARY

Pediatric head and neck tumors are uniquely challenging because of the diagnostic diversity, small patient anatomy, and potential risks to their long-term growth and development. Although most tumors of the pediatric head and neck are benign, all head and neck tumors deserve detailed attention. Longitudinal experience within a focused team will allow a more accurate understanding of true morbidity to inform comprehensive oncologic planning discussions. Multidisciplinary care with oncology, otolaryngology, reconstructive surgery, and oral surgery affords the greatest opportunity for the best oncologic, cosmetic, and functional outcomes.

CLINICS CARE POINTS

- Pediatric head and neck tumors are diagnostically diverse and require thoughtful multidisciplinary evaluation and treatment.
- Complex pediatric head and neck resection and reconstruction can be safe and effective with detailed consideration to oncologic, cosmetic and functional outcomes.

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