Reza Rahbar · Carlos Rodriguez-Galindo
John G. Meara · Edward R. Smith
Antonio R. Perez-Atayde *Editors*

Pediatric Head and Neck Tumors

A–Z Guide to Presentation and Multimodality Management

Springer

Pediatric Head and Neck Tumors

Reza Rahbar · Carlos Rodriguez-Galindo
John G. Meara · Edward R. Smith
Antonio R. Perez-Atayde
Editors

Pediatric Head and Neck Tumors

A-Z Guide to Presentation and Multimodality Management

Springer

Editors
Reza Rahbar DMD, MD
Department of Otolaryngology
Harvard Medical School Boston Children's Hospital
300 Longwood Avenue - LO367
Boston, MA, USA

John G. Meara
Harvard Medical School Boston Children's Hospital
Department of Plastic and Oral Surgery
300 Longwood Avenue Enders 114
Boston, MA, USA

Antonio R. Perez-Atayde
Harvard Medical School Boston Children's Hospital
Department of Pathology
300 Longwood Avenue
Boston, MA, USA

Carlos Rodriguez-Galindo
Dana-Farber/Boston Children's Cancer and Blood Disorders Center
Harvard Medical School
450 Brookline Avenue D-3133
Boston, MA, USA

Edward R. Smith
Harvard Medical School Boston Children's Hospital
300 Longwood Avenue
Boston, MA, USA

ISBN 978-1-4614-8754-8
ISBN 978-1-4614-8755-5 (eBook)
DOI 10.1007/978-1-4614-8755-5
Springer New York Heidelberg Dordrecht London

Library of Congress Control Number: 2013951830

Printed on acid-free paper

Springer is part of Springer Science+Business Media (www.springer.com)

Preface

Treatment of children with head and neck tumors exemplifies the complex multidisciplinary care that defines modern pediatric practice. From benign to malignant conditions, each tumor and each patient requires a dedicated team of specialists that understand the disease and define the best course of action, applying the most effective treatments that maximize cure options with minimal adverse effects. Accomplishing these goals requires seamless integration of many disciplines, including pathology, diagnostic and interventional radiology, otorhinolaryngology, skull base, plastic and ocular surgery, and pediatric and radiation oncology.

The Head and Neck Tumors Program at Boston Children's Hospital and Dana-Farber Cancer Institute was formed to provide a well-integrated team approach to children with those complex and often devastating diseases. In this book, we have invited a team of experts from our program to share their knowledge in the diagnosis and management of the tumors that we encounter in our practice. We would like this book to be an A to Z practical guide that provides concise reviews and treatment recommendations for the different tumors.

This work represents the efforts of many. We would like to thank all our colleagues who have so generously shared their expertise and their time, and the editors who have so patiently helped us in this process. Most of all, we are grateful to our patients and their families, whose courage and determination inspire us to continue to work, to learn, and to advance our knowledge in the treatment of these disorders.

Boston, MA, USA — Reza Rahbar, MD
Boston, MA, USA — Carlos Rodriguez-Galindo, MD
Boston, MA, USA — John G. Meara, MD, DMD, MBA
Boston, MA, USA — Edward R. Smith, MD
Boston, MA, USA — Antonio R. Perez-Atayde, MD, PhD

Contents

Contributors

Natasha M. Archer Department of Pediatric Hematology-Oncology, Dana-Farber/Boston Children's Cancer and Blood Disorders Center, Roxbury, MA, USA

Lissa C. Baird Department of Neurosurgery, Oregon Health & Science University, Portland, OR, USA

Jacob R. Brodsky Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Rafael A. Couto Department of Plastic and Oral Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Michael J. Cunningham Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Alfredo A. Dela Rosa Department of Oral and Maxillofacial Surgery, University of the Pacific, School of Dentistry, San Francisco, CA, USA

A. Lindsay Frazier Department of Pediatric Oncology, Dana-Farber Cancer Institute, Boston, MA, USA

Catherine M. Gordon Department of Adolescent Medicine, Hasbro Children's Hospital, Providence, RI, USA

Adam L. Green Pediatric Oncology, Dana-Farber Cancer Institute, Boston, MA, USA

Lillian M. Guenther Department of Medicine, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Stephen A. Huang Department of Medicine, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Katherine A. Janeway Department of Pediatric Oncology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Dwight T. Jones Department of Otolaryngology - Head and Neck Surgery, University of Nebraska College of Medicine, Omaha, NE, USA

Vikramjit S. Kanwar Pediatric Hematology-Oncology, Albany Medical Center, Albany, NY, USA

Stephen Kieran Department of Otolaryngology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Sang Yoon Kim Department of Oral and Maxillofacial Surgery, INOVA Fairfax Hospital, Vienna, VA, USA

Ilkka Kivekäs Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School Boston, MA, USA

Department of Otorhinolaryngology, Tampere University Hospital and the University of Tampere, Tampere, Finland

Brian Labow Department of Plastic and Oral Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Paul Lennon Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Jennifer W. Mack Dana-Farber Cancer Institute, Boston, MA, USA

Department of Pediatric Hematology/Oncology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Karen J. Marcus Pediatric Radiation Oncology, Dana Farber/Boston Children's Cancer and Blood Disorders Center, Brigham and Women's Hospital, Harvard Medical School, Boston, MA, USA

Trevor McGill Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Brendan McNeish Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

John G. Meara Department of Plastic and Oral Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Biren P. Modi Department of Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Allison O'Neill Department of Pediatric Oncology, Harvard Medical School, Dana-Farber/Boston Children's Hospital Cancer and Blood Disorders Center, Boston, MA, USA

Darren B. Orbach Department of Radiology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Interventional & Neurointerventional Radiology, Department of Radiology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Padua Jr. Department of Radiology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Horacio M. Padua Department of Radiology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Bonnie L. Padwa Department of Plastic and Oral Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Antonio R. Perez-Atayde Department of Pathology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Dennis Poe Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Sanjay P. Prabhu Department of Radiology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Mark Proctor Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Melissa S. Putman Department of Endocrinology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Reza Rahbar Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Carlos Rodriguez-Galindo Department of Pediatric Oncology, Dana-Farber/Boston Children's Hospital Cancer and Blood Disorders Center, Harvard Medical School, Boston, MA, USA

R. Michael Scott Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Raja Shaikh Department of Interventional Radiology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Robert C. Shamberger Department of Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Suzanne Shusterman Department of Pediatric Hematology-Oncology, Dana-Farber/Boston Children's Cancer and Blood Disorders Center, Boston, MA, USA

Edward R. Smith Department of Neurosurgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Lisa M. Stafford College of Nanoscale Engineering, Albany, NY, USA

Amir Taghinia Department of Plastic and Oral Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Behroze Adi Vachha Department of Radiology, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA, USA

Deborah Vanderveen Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Christian J. Vercler Department of Plastic and Oral Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Section of Plastic Surgery, University of Michigan, Ann Arbor, MI, USA

Frank W. Virgin Department of Otolaryngology Head and Neck Surgery, Monroe Carell Jr. Children's Hospital at Vanderbilt, Nashville, TN, USA

Mark S. Volk Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Lynda Vrooman Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Benjamin C. Warf Department of Neurosurgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Karen Watters Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Christopher Weldon Department of Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Kenneth R. Whittemore, Jr. Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

Part I
Evaluation and Management

Imaging

1

Behroze Adi Vachha and Sanjay P. Prabhu

Introduction

Head and neck masses are relatively common in children. Unlike adults, where the majority of neck lesions encountered are malignant, neck masses in children are usually (>90%) benign. Neck masses may be a result of a variety of congenital, infective, inflammatory, traumatic, lymphovascular, and neoplastic etiologies [1, 2].

Imaging assessment of neck masses is tailored based on the child's symptoms and findings on clinical exam. Goal of imaging should be to generate a limited list of differential diagnoses or in some cases, specify a single definitive diagnosis while keeping the ionizing radiation exposure as low as reasonably achievable (the ALARA principle).

If a lesion is thought to be neoplastic, further imaging is aimed at characterizing tumors and providing a more refined differential diagnosis, assessing the extent of the lesion, detecting involvement of adjacent structures and determining metastatic spread if the tumor is malignant, all of which are essential for appropriate treatment planning as well as to determine the prognosis of malignant tumors. Imaging is also used to guide needle biopsy and to follow response to therapy. It is important to note that while imaging can narrow the differential diagnosis of pediatric head and neck masses, biopsy and/or excision may still be required for definitive therapy.

Head and neck tumors are less common with only 5% of pediatric primary malignancies arising in the head and neck region [3]. Imaging plays an important role in the differentiation of the more benign entities from malignancies. Early diagnosis is critical as many pediatric head and neck malignancies are readily treatable and often curable by current medical and surgical management when detected early.

In this chapter we outline the various imaging techniques used to assess head and neck neoplasms (benign and malignant) in the pediatric population and review the imaging findings of most common pediatric benign and malignant tumors.

Overview of Imaging Techniques

A tailored multimodality imaging approach utilizing varying combinations of ultrasound (US), computed tomography (CT), magnetic resonance imaging (MRI), and radionuclide studies is useful in characterizing pediatric neck tumors and allows for appropriate management.

Ultrasound (US)

US is often the initial imaging modality of choice in the evaluation of palpable extracranial head and neck tumors and assessment of superficial glandular structures such as the thyroid and salivary glands in children [2].

Advantages of US in children include smaller neck size and relative lack of subcutaneous fat in children results in better sonographic penetration and resolution [4]. US plays an important role in distinguishing solid from cystic lesions and differentiating nodal from non-nodal masses [5]. In contrast to CT and MRI, US provides real-time, rapid noninvasive imaging at a lower cost and does not involve ionizing radiation exposure. It is portable and can be performed at the bedside without the need to sedate the child.

Drawbacks of US include the dependency on operator skill and experience and inherent lower spatial resolution and tissue contrast than cross-sectional imaging modalities. Optimizing US technique can help improve image quality and aid diagnosis.

S. P. Prabhu (✉)
Department of Radiology, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA
e-mail: sanjay.prabhu@childrens.harvard.edu

B. A. Vachha
Department of Radiology, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA, USA

R. Rahbar et al. (eds.), *Pediatric Head and Neck Tumors,*
DOI 10.1007/978-1-4614-8755-5_1, © Springer Science+Business Media New York 2014

Ideally, the patient is scanned in a supine position with the neck slightly hyperextended to optimize field of view. Using high frequency linear array transducers (7–12 MHz) to evaluate superficial neck structures, small footprint high frequency small part transducers in infants and curved or sector transducers (6–8 MHz) to provide improved resolution of deeper structures in the neck are among the methods to optimize US of the neck in children [3, 4]. Color Doppler examinations and spectral tracings should be used to evaluate presence of and pattern of vascular flow within the mass [5].

Computed Tomography (CT)

Advantages of CT include its ready availability in emergent settings and ability to detect osseous changes caused by the mass such as remodeling or erosion and intralesional calcification better than MRI and US. Multidetector CT (MDCT) scanners allow for rapid scan acquisition without compromising image quality. This is especially useful in critically ill children who cannot reliably suspend respiration. Also, fast scan times decrease the potential for motion degradation and may obviate the need for sedation. Volumetric 3-D reconstruction of lesions after MDCT acquisition can be used to plan surgical approaches and assess tumor response following treatment.

Main limitation of CT in pediatric populations includes risks of ionizing radiation exposure associated with CT, particularly the potential carcinogenic effects [4, 6–8]. Technical options are now included in newer CT scanners in an effort to reduce the dose from CT exams. These include x-ray beam filtration and collimation, tube current modulation tailored to patient size and indication, peak kilovoltage optimization, improved detector efficiency, and noise reduction algorithms [9, 10]. Adhering to the ALARA concept entails applying strategies that reduce radiation exposure to the child without compromising diagnostic accuracy and alternative methods of imaging like MRI and US should be explored in all cases [11]. CT has lower tissue contrast resolution compared to MRI.

Certain technical issues need to be considered to ensure that the maximum information is gained from the CT scan. For CT of the soft tissues of the neck, the child is usually placed in the supine position with the neck slightly extended to exclude the orbits. Most studies can be performed with the child breathing quietly. Region scanned usually extends from the skull base to the top of the aortic arch. Intravenous contrast should be administered if there are no contraindications to the use of contrast for better delineation of masses from adjacent structures and to determine tumor enhancement patterns. For contrast-enhanced studies, split bolus techniques (wherein half the contrast is administered and images are then obtained after 3-min pause during the administration of the second half of the contrast bolus) provide better lesion and vascular enhancement without the need for multiple phases of scanning, which increase the radiation dose [5]. Multiplanar reconstructions are generated from the initial data set to avoid repeated scans.

Magnetic Resonance Imaging (MRI)

MRI is the ideal modality of choice for investigating neck masses due to its superior soft tissue resolution and avoidance of ionizing radiation. Contrast-enhanced MRI better defines lesion extent and margins and it can detect perineural spread of tumor and intracranial extension. Although CT provides better illustration of subtle cortical erosion, bone infiltration and cartilage invasion by soft tissue lesions is detected earlier and defined better by MRI.

The main disadvantage of MRI in children is that the many sequences require the child to lie still for a substantially longer amount of time than that needed for a CT and therefore, sedation of younger children is often required to reduce motion artifacts. Artifacts after surgical reconstruction with metallic hardware limit visualization on MRI due to susceptibility artifact, particularly on images employing fat saturation and echoplanar imaging.

MRI technique and sequence selection should be optimized based on the age of the child, the location and type of neck mass being investigated. Patients older than age 6 are placed in the supine position with the neck slightly extended and the study is performed with the child breathing quietly. Infants may be fed prior to the exam and swaddled to minimize motion artifact (“feed and wrap”). Slightly older pediatric patients (less than 6 years) often require sedation to optimize image acquisition [4].

Indication-based protocols should be employed to ensure that the diagnosis is determined with the least number of sequences and within the shortest time possible. This approach helps minimize duration of sedation and avoids the risk of patient motion in younger patients being scanned without sedation. Most head and neck protocols include multiplanar T1, fat-suppressed T2 or STIR images, a flow-sensitive gradient echo sequence, and contrast-enhanced multiplanar fat-suppressed T1-weighted sequences. Sagittal imaging may be considered for lesions around the temporomandibular joint, tongue base, nasopharyngeal, and airway lesions [12].

Diffusion-weighted imaging (DWI) has shown some value in characterization of head and neck mass lesions in children. As a rule of thumb, malignant pediatric tumors have lower apparent diffusion coefficient (ADC) than that of benign solid and cystic lesions, likely reflecting increased lesion cellularity [13]. For example, rhabdomyosarcomas (RMSs)have the lowest ADC values and mucoepidermoid carcinomas have higher ADC values than sarcomas [13].

Additional studies are required to assess the value of DWI in initial diagnosis and evaluation of response-following therapy of pediatric head and neck neoplasms.

Radionuclide Studies (Positron Emission Tomography (PET) and PET-CT)

Unlike in adults, the role of PET in management of all pediatric solid tumors is less well-defined. However, ^{18}F fluorodeoxyglucose PET (FDG-PET) and FDG-PET-CT are important tools in the noninvasive evaluation, initial staging, and continued monitoring of children with certain types of malignancies (e.g., lymphomas and some sarcomas) [3, 14, 15].

Key advantage of FDG-PET-CT over MRI or CT is the ability to distinguish viable recurrent or residual tumor from post-therapeutic changes [14].

PET and PET-CT has the risks of ionizing radiation. Further, accurate anatomic coregistration of PET and CT images requires that the child remains still throughout the procedure. As these exams can be lengthy, younger patients often require sedation or, occasionally, general anesthesia to avoid misregistration [14, 15].

Physiologic variations in FDG distribution in children include higher uptake of FDG in thymus, adenoids, and tonsils, within metabolically active brown adipose tissue, bone marrow, and spleen [16, 17]. Uptake in the bone marrow and spleen may falsely suggest metastatic disease [14, 18, 19]. Additionally, intense FDG activity in brown adipose tissue can potentially mask cervical, supraclavicular, and axillary pathology in pediatric patients [15].

Despite these limitations, PET-CT holds promise as an alternative response of assessing tumor response to therapy.

Differential Diagnosis

The following section deals with tumors, but it is important to note that infective, inflammatory, and lymphovascular lesions are relatively more common in a child and should be considered in the differential diagnosis of head and neck masses.

Specific Tumor Types

As clinical and pathological aspects of individual tumors are dealt with elsewhere in this book, the following discussion focuses on imaging characteristics of the more common head and neck benign and malignant tumors seen in the pediatric age group.

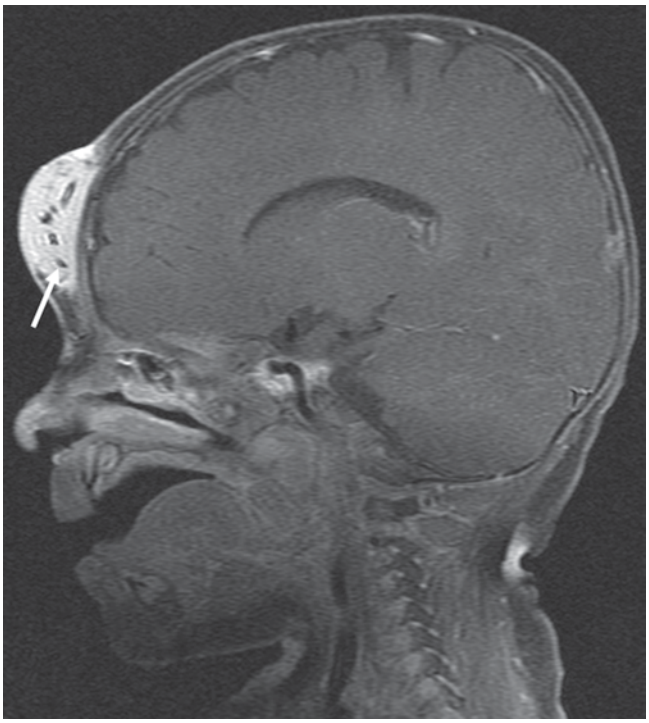

Fig. 1.1 Hemangioma. Post contrast sagittal fat-suppressed T1W image in a 6 month-old male demonstrates a large midline subcutaneous hemangioma over the forehead, characterized by marked enhancement and prominent signal void indicating presence of blood vessels (*arrow*)

Benign Tumors

Hemangioma

Hemangioma is the most common vascular tumor and arises in infants. They proliferate rapidly during the first year of life and involute over the next few years.

During the proliferative phase, US and color Doppler examinations demonstrate a soft tissue mass with prominent vessels and arterial and venous waveforms. Peak venous velocities are not as high as seen in a true arteriovenous malformation (AVM). During the involutional stage, increasing fibrofatty tissue is seen within the lesion.

Contrast-enhanced CT (CECT) demonstrates a soft tissue lobulated mass with diffuse contrast-enhancement and prominent vessels in and adjacent to the mass (Fig. 1.1).

On MRI, the lesion appears isointense to muscle during the proliferative phase and demonstrates fatty replacement during the involutional phase on T1-weighted sequences. It is mildly hyperintense to muscle on T2-weighted images. Fat saturated T1-weighted contrast-enhanced sequences demonstrate intense contrast enhancement with serpiginous flow voids in and adjacent to the mass.

Note should be made of associated abnormalities in the brain and chest in view of the known association of **p**osterior fossa malformations, **h**emangiomas, **a**rterial anomalies, **c**oarctation of the aorta and **c**ardiac defects, **e**ye abnormalities,

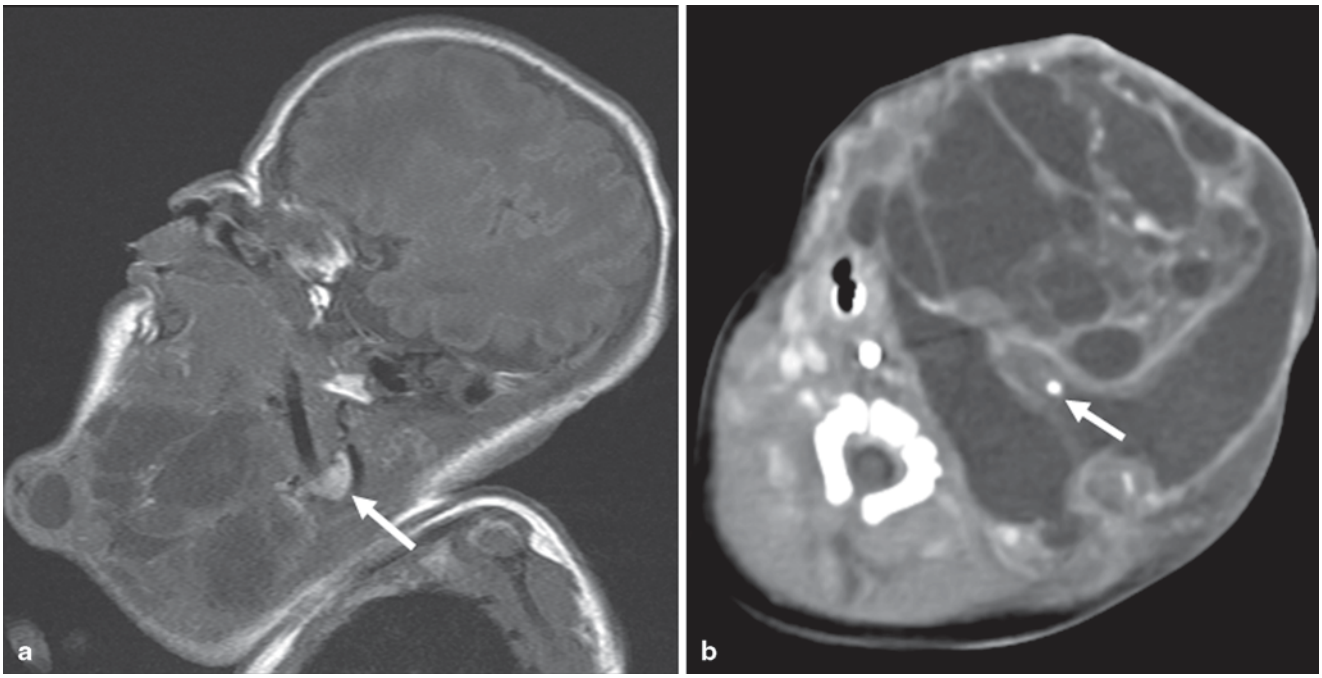

Fig. 1.2 Teratoma. **a** Sagittal T1W image shows a large mixed solid and cystic cervicofacial mass in a newborn infant. Note the hyperintense structure in the neck is the right lobe of the thyroid (*arrow*). The mass involved the left lobe of the thyroid. **b** Axial CECT shows scattered calfications within the lesion (*arrrow*)

sternal malformations, and **s**upraumbilical raphe (PHACES syndrome).

Differential diagnoses of hemangiomas include slow flow vascular malformations (venous and lymphatic malformations), arteriovenous malformations, plexiform neurofibroma, and sarcoma.

Teratoma

Teratomas are the commonest congenital head and neck tumors. Some cervicofacial teratomas are being increasingly diagnosed on antenatal US and/or MRI. These lesions present as large cervical masses can cause fatal airway compression at birth.

US demonstrates a predominantly solid or mixed cystic/solid structure.

Calcifications are virtually pathognomonic of teratoma but are seen in only half the cases and are better delineated by CT. CT demonstrates a heterogenous mass with areas of fat attenuation and calcification.

MRI signal intensities are variable and depend on the internal composition of the lesion. Presence of fat can be confirmed by using fat-saturated images (Fig. 1.2).

Differential diagnoses of cystic teratomas include lymphatic malformations and rarely infantile myofibromatosis. A useful imaging differentiating feature is that involvement of the thyroid gland by an infrahyoid congenital mass is almost pathognomonic of a teratoma (considered by some authors to be arising from the thyroid) [20].

Nerve Sheath Tumors

Plexiform neurofibromas are benign peripheral nerve sheath tumors, virtually diagnostic of neurofibromatosis Type 1. Extracranial head and neck plexiform neurofibromas arise most commonly from the trigeminal nerve at the orbital apex [21]. These lesions present as multiple masses or as fusiform enlargement of the peripheral nerves produce a "bag-of-worms" appearance [22]. On MRI, these lesions are typically hyperintense on T2-weighted images and hypointense on T1-weighted images (Fig. 1.3). Deeper lesions are typically nodular and superficial lesions have a more diffuse, infiltrating appearance involving the subcutaneous tissues and the skin.

Juvenile Nasopharyngeal Angiofibroma (JNA)

Imaging (either CT or MRI) usually confirms the diagnosis of JNA and in almost all cases, should help avoid biopsy. CT and MRI are utilized for presurgical planning of JNAs. MRI enables assessment of soft tissue extent and CT to determine the presence of skull base erosion.

Ideal imaging protocols for preoperative planning and staging include a maxillofacial CT with multiplanar reformats, maxillofacial MRI with T1-weighted fat-saturated contrast-enhanced sequences and catheter angiography of the external and internal carotid arteries (ECA and ICA, respectively) to identify feeding vessels. MR angiogram (MRA) may be performed to help evaluate the need for, and when needed, help plan catheter angiography for presurgical embolization.

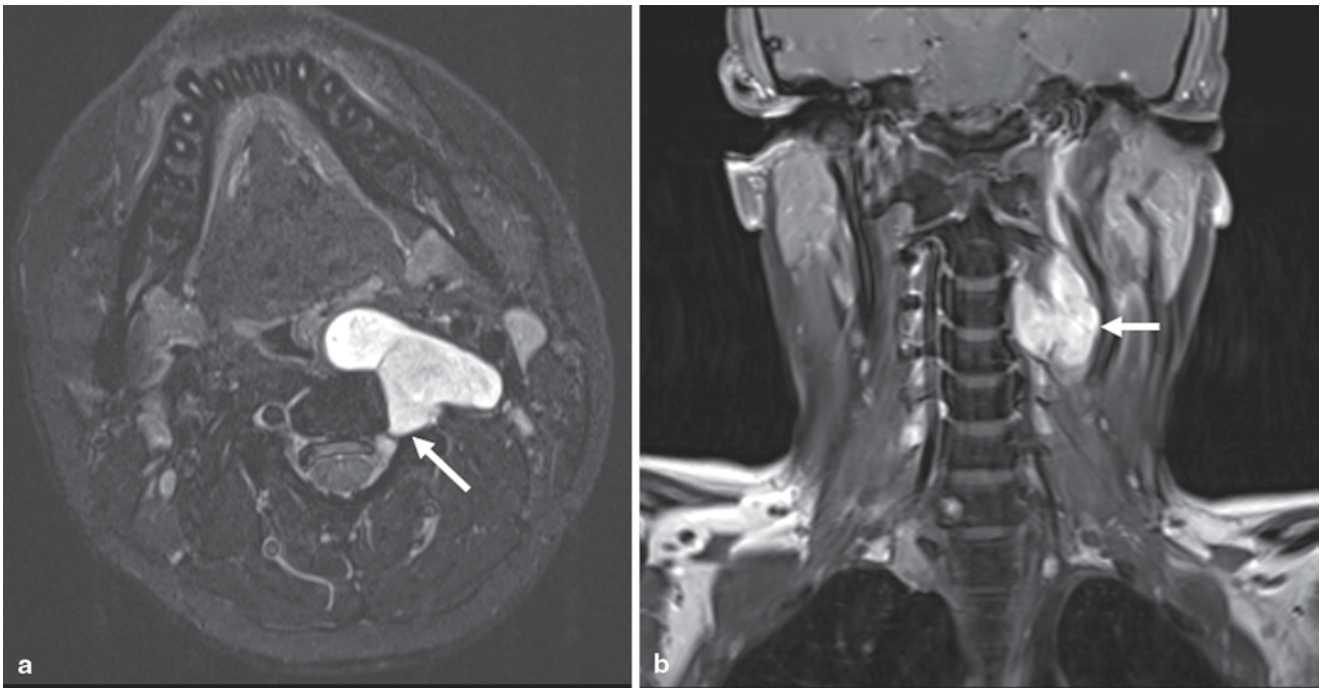

Fig. 1.3 Neurofibroma. **a** Axial fast spin echo inversion recovery (FSEIR) image shows a well-circumscribed lobulated T2 hyperintense lesion arising from the left C3-4 neural foramen in a 16 year-old male with neurofibromatosis Type 1 (*arrow*). **b** The lesion enhances avidly following contrast as seen on the coronal T1W fat-saturated images (*arrow*)

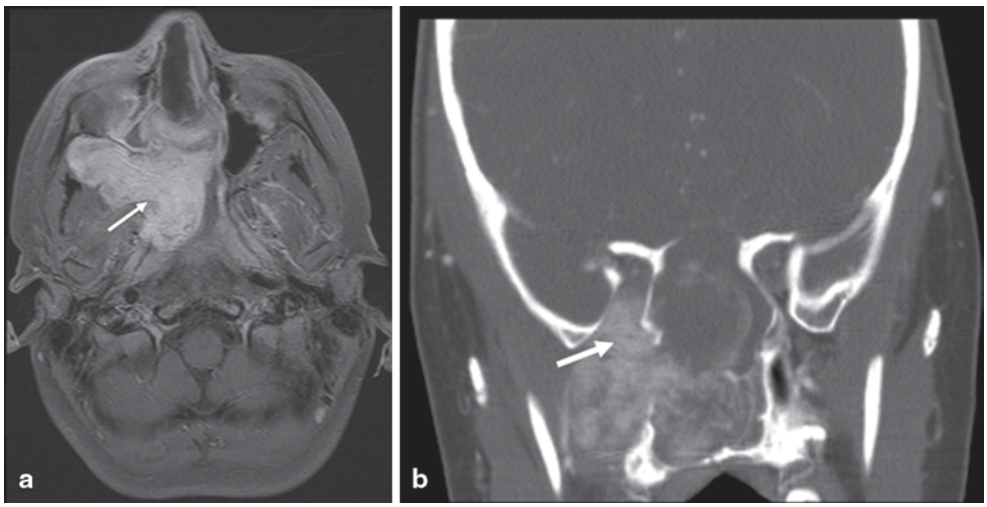

Fig. 1.4 JNA. **a** Axial fat-suppressed post contrast T1W image shows a intensely enhancing nasopharyngeal mass, eroding the sphenoid and ethmoid sinuses, expanding the pteryogopalatine fossa, and extending through the pterygomaxillary fissure into the infratemporal fossa (*arrow*) in a 12-year-old male presenting with epistaxis. **b** Coronal reformat of contrast-enhanced CT shows extension of the mass into the foramen rotundum and inferior orbital fissure (*arrow*) and extensive osseus destruction

CT usually reveals a diffusely enhancing soft tissue mass arising at the sphenopalatine foramen and extending from the posterior nasal cavity into the nasal cavity, nasopharynx, and pterygopalatine fossa (Fig. 1.4). Widening of the ipsilateral nasal cavity and pterygopalatine fossa and bowing of posterior wall of the maxillary sinus anteriorly is noted.

On MRI, a heterogeneous mass with intermediate signal is seen on T1- and T2-weighted sequences. Serpentine flow voids are typically seen within the tumor with intense enhancement post contrast administration. Coronal T1-weighted images are required to look for cavernous sinus, sphenoid sinus, and skull base extension.

Catheter angiography demonstrates a capillary blush fed by feeding vessels (usually ascending pharyngeal or internal maxillary arteries) from the ECA or occasionally, in the case of skull base or cavernous sinus extension, from the ICA.

Differential diagnoses include antrochoanal polyp, RMS, and hemangioma.

Langerhans Cell Histiocytosis (LCH)

LCH is typically characterized on CT by an enhancing soft tissue mass associated with bony involvement, which classically involves "punched out" lytic lesions. However, bony lesions may also present with irregular sclerotic margins or fragments of bone associated with smaller or no appreciable soft tissue component.

On MRI, LCH lesions show ill-defined borders, which are iso- to hypointense on T1-weighted and iso- to hyperintense on T2-weighted sequences with homogenous enhancement. The demonstration of enhancing masses on MRI helps distinguish LCH occurring in the temporal bone from other erosive processes of the temporal bone [5].

Differential diagnoses include acquired cholesteatoma, cholesterol granuloma, acute mastoiditis, and RMS.

Malignant Tumors

The most common pediatric head and neck malignancies include lymphomas, RMSs, thyroid malignancies, nasopharyngeal carcinomas (NPCs), salivary gland malignancies, neuroblastomas, and malignant teratomas.

Hodgkin Lymphoma (HL) and Non-Hodgkin Lymphomas (NHL)

US may be used to assess superficial cervical lymph nodes. Sonographic features of malignancy include increased size, loss of the normal oval shape with a more round shape, and loss of the normal echogenic hilum [4, 5]. Doppler US may demonstrate displacement of vessels, subcapsular vessels or aberrant vessels, or avascular foci.

CECT is the imaging modality of choice in assessing the disease and extent of extranodal spread, particularly involvement of lungs. CECT should include the neck chest, abdomen, and pelvis for accurate staging and may be coregistered with PET scans. Oral contrast is administered prior to the scan to help evaluate abdominal disease burden optimally.

Variable enhancement of lymph nodes may be noted (Fig. 1.5). Lymph nodes measuring less than 1 cm in short axis diameter are usually considered normal by size criteria. Central hypodensity may indicate nodal necrosis. If there is lack of fat stranding and less intense enhancement, consider lymphoma instead of infectious lymphadenitis [5]. Burkitt's lymphoma may be seen on CT as a soft tissue mass with bony involvement of the mandible and "floating teeth" [5].

MRI demonstrates enlarged, round nodes which are isointense to hypointense to muscle on T1-weighted sequences, mildly hyperintense on T2-weighted sequences, and with less avid enhancement than reactive lymph nodes following the administration of gadolinium.

FDG-PET has been shown to be superior to Gallium 67 scans in staging, evaluating tumor response to therapy and determining tumor relapse [23].

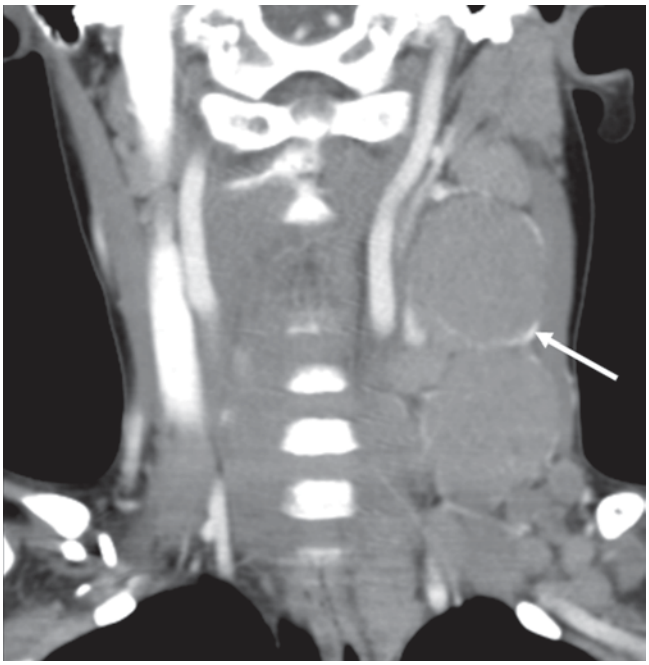

Fig. 1.5 HL. Coronal reformat of CECT shows large rounded cervical chain lymph nodes with peripheral enhancement (*arrow*)

Rhabdomyosarcoma (RMS)

RMSs are typically bone-destroying and "bone-pushing" tumors. CT best depicts this osseous change. Both CT and MRI demonstrate a soft tissue mass with variable enhancement. RMS is iso- to hypointense to muscle on T1-weighted sequences and hyperintense to muscle on T2-weighted sequences with moderate-to-intense enhancement following contrast administration (Fig. 1.6) Fat-suppressed T1-weighted images are helpful for the detection of orbital masses and parameningeal tumors. MRI also helps delineate intracranial extension of parameningeal RMSs.

Follow-up imaging using the same imaging modality should be performed no earlier than 6 weeks post therapy to avoid confusion between post-therapeutic change and residual disease. Enhancement of the tumor bed after 6 weeks after therapy is considered suspicious for recurrent or residual tumor [4].

The differential diagnosis on imaging varies according to the location of the tumor and includes lymphoma, nasopharyngeal carcinoma, metastatic neuroblastoma, JNA, and LCH.

Thyroid Malignancies

Approximately 2% of all thyroid cancers occur in children and adolescents. When a solitary thyroid nodule is identified in children and adolescents, approximately 20% of the lesions represent malignancy compared with 5% in adults [24]. Following initial measurement of serum thyroid-stimulating hormone (TSH), calcitonin (for diagnosis of medullary thyroid carcinoma), a neck US is the imaging modality of

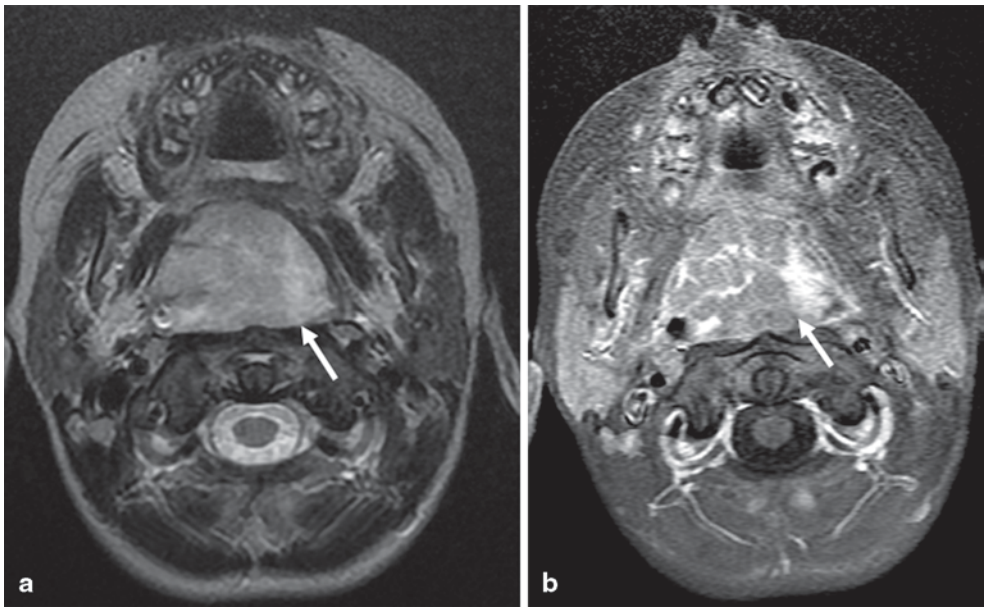

Fig. 1.6 Nasopharyngeal RMS. **a** Axial T2W image shows a large well-defined slightly T2-hypointense mass in the nasopharynx in a 28-month-old male (*arrow*). **b** Axial fat-suppressed post contrast T1W image shows the mass enhances heterogenously (*arrow*)

choice. Sonographic features suggestive of malignancy include ill-defined margins, microcalcifications, and variable echogenicity. Fine needle aspiration (FNA), which may be performed with or without US guidance may be useful for distinguishing benign and malignant nodules, but data are limited in children.

Metastases to regional cervical lymph nodes are most common in papillary thyroid carcinoma and occur in up to 90% of children affected by this type of thyroid malignancy [25].

Nasopharyngeal Carcinoma (NPC)

NPC is rare in pediatric populations and accounts for about 5% of pediatric head and neck malignancies. Children have greater bulk disease at presentation with relatively higher involvement of cranial nerves, lymph nodes, and skull base [4, 5].

NPC is characteristically seen on imaging studies as an asymmetric mass arising in the Fossa of Rosenmuller. CECT demonstrates a homogenously enhancing soft tissue mass centered in the lateral pharyngeal recess of the nasopharynx commonly associated with cervical adenopathy and skull base erosion.

On MRI, the mass is iso- to hypointense to muscle on T1-weighted sequences and hyperintense on T2-weighted sequences with homogenous enhancement following contrast administration (Fig. 1.7). Coronal contrast-enhanced T1-weighted images best depict intracranial extension of the tumor through skull base foramina. PET-CT shows FDG avid nodes.

Cervical lymph node involvement is present in 80–90% of patients at presentation, 50% of which are bilateral. As opposed to NPC in adults, necrosis within metastatic lymph nodes is uncommon in children.

Differential diagnosis based on location includes lymphoma, benign-mixed tumor, minor salivary gland malignancy, and lymphoid hyperplasia.

Salivary Gland Tumors

Although primary tumors of the salivary glands are uncommon in children, the ratio of malignant tumors to benign lesions is slightly higher in children than in adults. Tumors most commonly arise in the parotid glands. The commonest primary malignancy is mucoepidermoid carcinoma. The salivary glands may also be involved as an extra nodal site in NHL.

US, CT, and MRI are used for the evaluation of salivary gland lesions. US helps assess the size of the gland, distinguish diffuse from focal disease, and assess vascularity within the lesion and the adjacent structures, and also differentiate cystic from solid lesions. Fine-needle aspiration may be performed under US guidance.

CT is the imaging test of choice if an inflammatory mass is considered more likely and is helpful to assess for presence of calcification. MRI helps define the margins of a salivary gland mass better than CT.

Benign salivary gland tumors have a well-defined outline and do not enhance avidly on post contrast images. Calcifications within a mass on CT are highly suggestive of a benign-mixed tumor (pleomorphic adenomas). Large tumors are often lobulated. Warthin tumors are seen as well-encapsulated, homogenous cystic, or solid lesions on MRI, often in the tail of the parotid gland.

Mucoepidermoid carcinoma is the commonest malignant tumor of the salivary gland in children. CT and MRI appearances of these tumors vary with tumor grade. Lower-grade lesions resemble a pleomorphic adenoma, whereas higher-

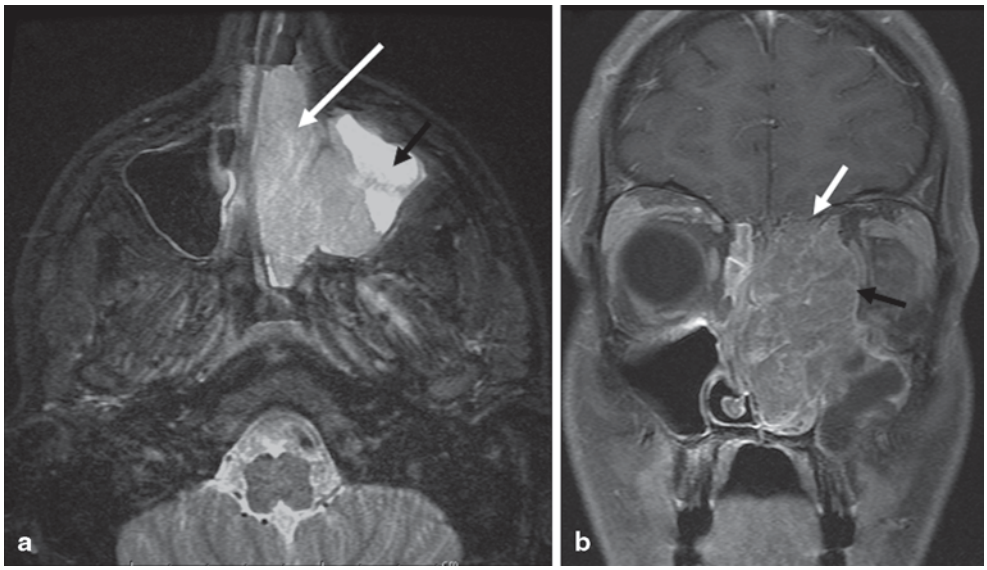

Fig. 1.7 NPC. **a** Axial FSEIR image shows sinonasal mass (*arrow*) centered along the medial left maxillary antrum and ethmoids, with low to intermediate T2 signal intensity in a 15-year-old male. Note the T2-hyperintense trapped secretions in the lateral aspect of the left maxillary sinus (*black arrow*). **b** Fat-suppressed post contrast coronal T1W image shows heterogeneous enhancement and parameningeal intracranial extension (*arrow*) through the left cribriform pate and ethmoid and also into the left orbit (*black arrow*)

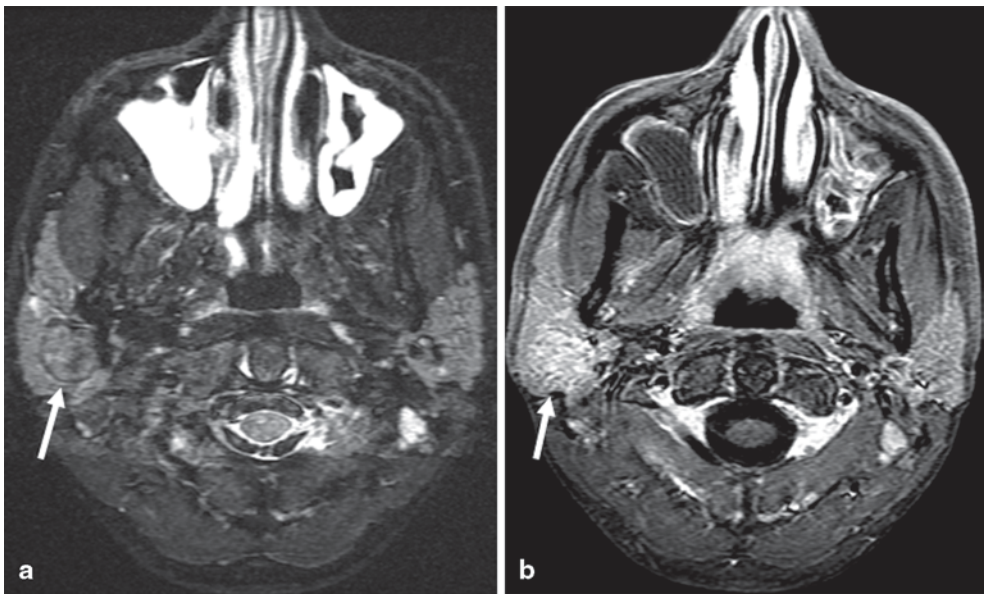

Fig. 1.8 Mucoepidermoid carcinoma. **a** Axial FSEIR image shows a well-defined heterogeneous mass (*arrow*) in the superficial and deep aspects of the right parotid gland. **b** Avid enhancement of the lesion is seen on axial fat-suppressed post contrast T1W image (*arrow*)

grade lesions have ill-defined, infiltrating margins and are more homogenous with variable enhancement (Fig. 1.8).

Neuroblastoma

Primary pediatric head and neck neuroblastomas are rare, with metastatic disease being the more common mode of involvement in this anatomic region.

Calcification may be seen on CT, but is less common in cervical neuroblastomas compared to abdominal neuroblastomas [4, 5]. Heterogeneous enhancement of the soft tissue mass is noted with CECT. Expansion of the diploic space due to marrow involvement and periosteal reaction is often seen (Fig. 1.9).

On MRI, the mass demonstrates hyperintense signal on T2-weighted images and shows heterogenous enhancement following the administration of gadolinium. Metaiodobenzylguanidine (MIBG) scans are used to assess bone and marrow involvement and in monitoring response to therapy [4].

Metastasis

Metastases in the head and neck occur more commonly to the osseous skeleton in children. These are present on CT as lytic and permeative lesions often with periosteal reaction and associated soft tissue masses. Cervical lymph nodes are variably involved. Neuroblastoma is the most common primary in children less than 2 years of age. Leukemic infiltrates are commoner in older children. Metastasis from sarcomas and other tumors is present as solitary or multiple masses. On MRI, these lesions are hypointense on T2-weighted images with avid enhancement on post contrast images.

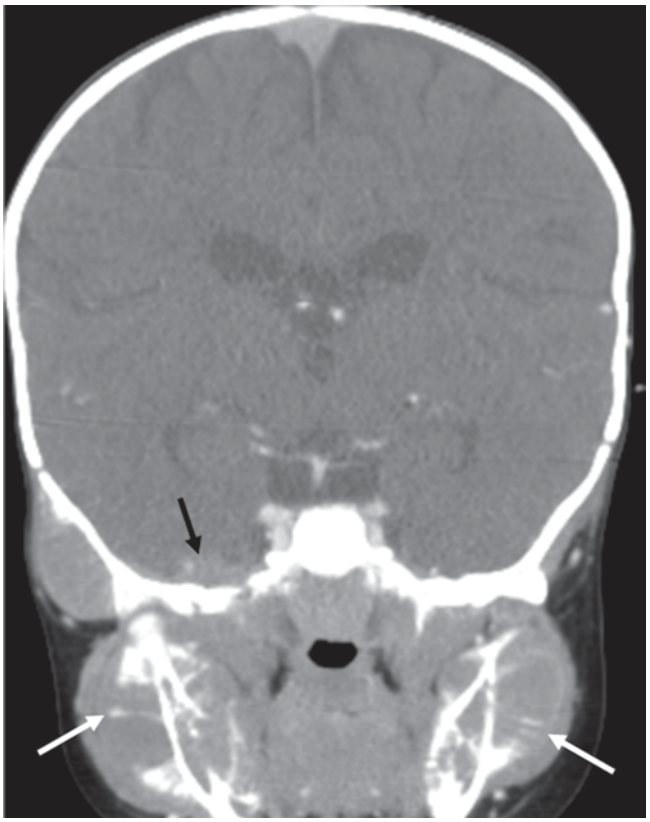

Fig. 1.9 Neuroblastoma. Coronal reformat CECT demonstrates multiple soft tissue masses around the face and neck with spiculated periosteal reaction in a 11-month-old female with increasing facial swelling and bilateral periorbital swelling and anemia (*arrows*). Note the intracranial involvement (*black arrow*)

Conclusion

The role of imaging has become increasingly important in providing maximum diagnostic information in preoperative/medical treatment planning and prognosis, and later in monitoring efficacy of therapy and detecting tumor recurrence. A carefully tailored multimodality imaging approach combined with careful history and clinical examination can help formulate a fairly accurate diagnosis and help direct appropriate patient management.

References

1. Turkington JR, Paterson A, Sweeney LE, Thornbury GD. Neck masses in children. Br J Radiol. 2005 Jan;78(925):75–85.
2. Friedman ER, John SD. Imaging of pediatric neck masses. Radiol Clin North Am. 2011 Jul;49(4):617–32, v.
3. Lloyd C, McHugh K. The role of radiology in head and neck tumours in children. Cancer Imaging. 2010;10:49–61.
4. Fefferman NR, Milla S. Ultrasound imaging of the neck in children. Ultrasound Clin. 2009;4(4):553–69.
5. Robson CD. Imaging of head and neck neoplasms in children. Pediatr Radiol. 2010 Apr;40(4):499–509.
6. Lauer MS. Elements of danger—the case of medical imaging. N Engl J Med. 2009 Aug 27;361(9):841–3.
7. Amis ES Jr, Butler PF. ACR white paper on radiation dose in medicine: three years later. J Am Coll Radiol. 2010 Nov;7(11):865–70.
8. Amis ES Jr, Butler PF, Applegate KE, et al. American college of radiology white paper on radiation dose in medicine. J Am Coll Radiol. 2007 May;4(5):272–84.
9. Mayo-Smith W. Protocol design. Image Wisely. 2010. p. 1–3.
10. Strauss KJ, Goske MJ, Kaste SC, et al. Image gently: ten steps you can take to optimize image quality and lower CT dose for pediatric patients. AJR Am J Roentgenol. 2010 Apr;194(4):868–73.
11. American College of Radiology. ACR-ASNR-SPR Practice guideline for the performance of computed tomography (CT) of the extracranial head and neck in adults and children. 2011.
12. American College of Radiology. ACR-ASNR Practice guideline for the perfomance of magnetic resonance imaging (MRI) of the head and neck. 2007.
13. Abdel Razek AA, Gaballa G, Elhawarey G, Megahed AS, Hafez M, Nada N. Characterization of pediatric head and neck masses with diffusion-weighted MR imaging. Eur Radiol. 2009 Jan;19(1):201–8.
14. Jadvar H, Connolly LP, Fahey FH, Shulkin BL. PET and PET/CT in pediatric oncology. Semin Nucl Med. 2007 Sep;37(5):316–31.
15. McCarville MB. PET-CT imaging in pediatric oncology. Cancer Imaging. 2009;9:35–43.
16. Patel PM, Alibazoglu H, Ali A, Fordham E, LaMonica G. Normal thymic uptake of FDG on PET imaging. Clin Nucl Med. 1996 Oct;21(10):772–5.
17. Weinblatt ME, Zanzi I, Belakhlef A, Babchyck B, Kochen J. False-positive FDG-PET imaging of the thymus of a child with Hodgkin's disease. J Nucl Med. 1997 Jun;38(6):888–90.
18. Hollinger EF, Alibazoglu H, Ali A, Green A, Lamonica G. Hematopoietic cytokine-mediated FDG uptake simulates the appearance of diffuse metastatic disease on whole-body PET imaging. Clin Nucl Med. 1998 Feb;23(2):93–8.
19. Sugawara Y, Fisher SJ, Zasadny KR, Kison PV, Baker LH, Wahl RL. Preclinical and clinical studies of bone marrow uptake of fluorine-1-fluorodeoxyglucose with or without granulocyte colony-stimulating factor during chemotherapy. J Clin Oncol. 1998 Jan;16(1):173–80.
20. Riedlinger WF, Lack EE, Robson CD, Rahbar R, Nose V. Primary thyroid teratomas in children: a report of 11 cases with a proposal of criteria for their diagnosis. Am J Surg Pathol. 2005 May;29(5):700–6.
21. Khanna G, Sato Y, Smith RJ, Bauman NM, Nerad J. Causes of facial swelling in pediatric patients: correlation of clinical and radiologic findings. Radiographics. 2006 Jan–Feb;26(1):157–71.
22. Lin J, Martel W. Cross-sectional imaging of peripheral nerve sheath tumors: characteristic signs on CT, MR imaging, and sonography. AJR Am J Roentgenol. 2001 Jan;176(1):75–82.
23. Hudson MM, Krasin MJ, Kaste SC. PET imaging in pediatric Hodgkin's lymphoma. Pediatr Radiol. 2004 Mar;34(3):190–8.
24. Dinauer CA, Breuer C, Rivkees SA. Differentiated thyroid cancer in children: diagnosis and management. Curr Opin Oncol. 2008 Jan;20(1):59–65.
25. Dähnert W. Radiology review manual. 7th ed. Philadelphia: Wolters Kluwer/Lippincott Williams Wilkins. 2011.

Reconstructive Surgery

2

Brian Labow and Amir Taghinia

Introduction

This chapter deals with the broad subject of reconstructive surgery in the management of pediatric head and neck tumors. The nature of the subject mandates a somewhat different format in that a wide array of tumor types and anatomic locations are considered. As such, an overview of the thought processes and management principles that guide the reconstructive surgeon will be outlined. Preoperative planning, intraoperative management, and specialized areas for reconstruction will be emphasized. Some details for specific defects and commonly used flaps and techniques will also be presented.

A few important caveats should also be stated at the outset. Many of the tumor types and resultant defects found in pediatric head and neck oncology are rare, and in some cases represent unique situations. As such, reconstructive treatment recommendations are rarely evidence-based and depend more on principles and experience rather than established protocols or algorithms. The literature supporting a given reconstructive modality is often quite limited, especially in pediatric patients and prospective well-controlled studies are lacking. The authors recognize that there is always more than one reconstructive option and that the patient's, parents', and surgeon's familiarity and comfort with the risk and rewards of various approaches may also play a role in determining the type of reconstruction method that is selected. As such, the material presented below should be viewed as a guide rather than a series of definitive treatment recommendations.

Preoperative Planning: General Considerations

Successful reconstruction of the pediatric head and neck invariably begins with careful preparation [1]. The reconstructive surgeon should be engaged as soon as it is determined that some form of reconstruction may be needed. Ideally, this should occur well in advance of tumor extirpation. This allows for a complete understanding of the diagnosis, adjuvant treatments and prognosis, as well as interdisciplinary communication by all treatment teams, including radiology. In particular, the reconstructive surgeon should be aware of what anatomic structures are definitely, likely, or possibly involved. Will immediate reconstruction be required? How will surgical margins be assessed? How likely is tumor involvement at the margins and will this mandate reexcision? What is the likelihood of local recurrence and subsequent resection? These questions should be openly discussed as the answers to these questions may influence the type and timing of reconstruction.

Adjuvant therapy and its timing should also be discussed. Radiation can significantly affect the choice of reconstructive procedure. When administered prior to resection and reconstruction, radiation can cause local tissues to be edematous and microcirculation poor [1, 2]. In this setting, local tissue rearrangement or local flaps may have a higher rate of failure. Conversely, radiation after reconstruction can produce long-lasting deleterious changes that may lead the reconstructive surgeon to defer certain elements of the reconstruction until later in childhood to avoid the direct effect of radiation on the reconstructed element in question (Fig. 2.1). In some instances, neoadjuvant chemotherapy may severely lower the ability of the patient to tolerate prolonged reconstructive procedures such as free-tissue transfers and necessitate less invasive procedures. In other cases, delays in wound healing from reconstructive complications can dangerously delay postoperative chemotherapy. In these instances, less complex reconstructive choices may be necessary initially to increase the likelihood of early, uncomplicated wound closure.

Once the reconstructive surgeon fully understands the anatomic requirements and other treatment modalities to be employed in management of the tumor, a series of reconstructive options should be generated. In some instances there may be one clear "first option", in other instances there

B. Labow (✉) · A. Taghinia
Department of Plastic and Oral Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA
e-mail: brian.labow@childrens.harvard.edu

R. Rahbar et al. (eds.), *Pediatric Head and Neck Tumors,*
DOI 10.1007/978-1-4614-8755-5_2, © Springer Science+Business Media New York 2014

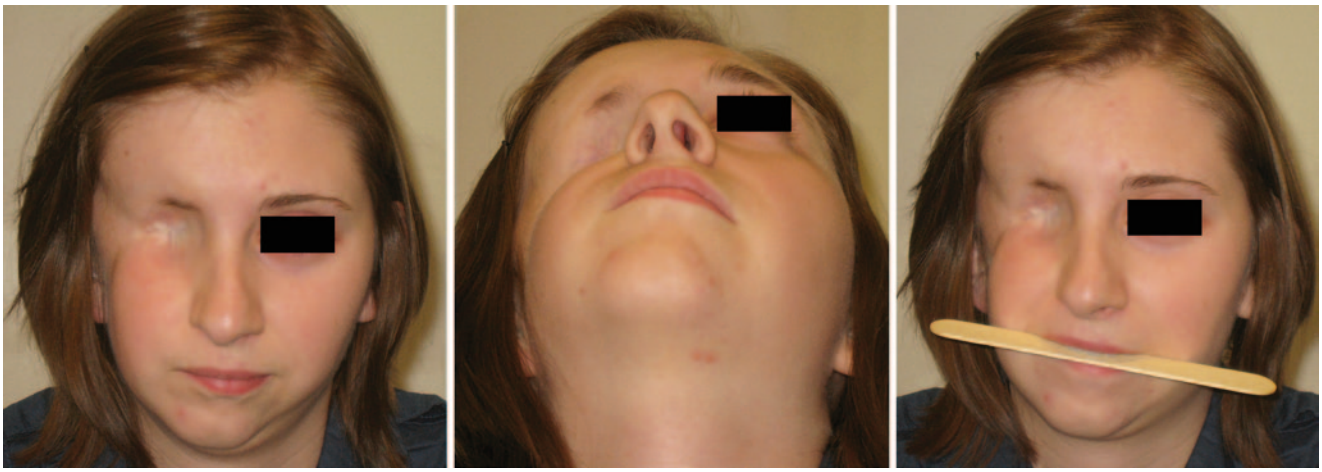

Fig. 2.1 Radiation effect. This adolescent patient underwent orbital extenteration for a rhabdomyosarcoma at the age of 4. She had free tissue transfer elsewhere followed by radiation. This case demonstrates the dramatic ill effects of radiation therapy on the growing maxillofacial skeleton. The mandible, maxilla, and orbit are substantially underdeveloped on the affected side

may be two or three equivocal options. Regardless, it is necessary to have at least one alternative procedure going into the operating room. This "lifeboat" may be deployed when intraoperative conditions change (e.g., unrecognized tumor progression, patient instability) or if the primary reconstruction modality is unsuccessful (e.g., partial or complete flap loss). When the reconstructive surgeon meets the patient and family, the rationale for the various the options should be fully discussed along with the advantages and disadvantages inherent to all reconstruction choices.

Equally important to interprovider consultation, preoperative planning must involve the parents and, when appropriate, the patient as well. The family will be overwhelmed by the diagnosis and there is often a sense of urgency to proceed as quickly as possible. The family may have been told that some form of "plastic surgery" or "reconstruction" will be required prior to the consultation with the reconstructive surgeon. A fine line must be walked between giving the family hope and inadvertently leading the family to have unrealistic expectations for the reconstruction. In addition to defining the defect and the reconstruction needs of the patient, the preferred treatment option(s) will be outlined. These may change based on anatomic considerations following physical examination or psychosocial considerations. For example, scarring from previous surgery may preclude specific donor sites for tissue or recipient vessels in case a microvascular procedure is required. Fortunately, unlike adult head and neck cancer patients, the effects of tobacco, diabetes, and other chronic comorbidities are rarely encountered. However, psychosocial considerations especially in adolescent patients, must be accounted for. It is important for the reconstructive surgeon to assess the family's and patient's understanding and tolerance for the reconstructive procedure being considered. In some cases, a simpler reconstruction with a less than ideal aesthetic outcome may be preferred if the surgical risks, recovery time, or postoperative restrictions are unacceptable to the patient or family.

All donor sites or potential donor sites for tissue, areas of scarring, and secondary deformities should be disclosed along with expectations for functional and aesthetic limitations at both the donor and recipient sites following surgery. Furthermore, depending on the age of the child, special attention should be given to the effects of growth on both of these locations. In many instances, additional procedures later in childhood will be required to address growth differences in the area of reconstruction. When this can be anticipated, the family should be made fully aware of a secondary procedure. In some instances, optimal reconstruction may require a series of staged procedures over time. Each patient and family should be viewed as unique with specific anatomic, psychological and social considerations. Care by the reconstructive team should be viewed as individualized, long-term, and may even exceed that of all other care team members.

Intraoperative Considerations

Timing and Sequence

A two-team approach is often helpful to minimize patient anesthesia and surgeon fatigue. In these situations, the free tissue flap is raised simultaneously with the extirpative operation. Clear communication between the oncologic and reconstructive teams is vital in these cases, especially when a

skin flap is required. With poor communication between the teams, it is not uncommon to raise a flap that is too small for the defect. Certainly, the safest approach is to wait until the defect is complete. In our experience, however, most cases are amenable to a two-team approach.

Anesthesia

If combined with cancer ablative operations, head and neck reconstructive procedures are often lengthy. An experienced anesthesia team is crucial for optimizing care and minimizing complications.

Airway In cases that involve the oropharynx, a nasal ray endotracheal tube is obligatory. The tube can be secured to the caudal septum with a heavy silk stitch. To avoid alar rim skin necrosis, the entire tubing apparatus should be brought inferiorly and secured to the patient's foam-padded forehead with tape. A straight accordion tube extender is often useful to lengthen the circuit and avoid kinks. The tubing closer to the anesthesia machine can also be secured to the back of the headrest for additional security. Once this process is complete, the surgeon should check the integrity of this construct by turning the head in either direction.

Positioning The positioning of the patient will depend to some extent on the reconstructive plan. In the case of pedicled flaps and most free tissue flaps, supine positioning is adequate. If a large defect is anticipated and a latissimus flap is considered for reconstruction, it may be prudent to harvest the muscle flap first in a lateral decubitus position, then partially close the donor site and turn the patient supine for the extirpative operation.

Tubes and Lines Hemodynamic instability is rare during resection and reconstruction of most pediatric head and neck tumors. The main exception to this is in large vascular malformations, especially arteriovenous malformations. As such, invasive monitoring is typically limited to an arterial line and at least one and usually two peripheral intravenous lines. If postoperative chemotherapy or frequent blood sampling is anticipated postoperatively, a central venous catheter may be placed at the outset of the procedure. In patients coming to the operating room with a previously placed port-a-cath™ or long-term indwelling central venous catheter, special care must be taken to ensure appropriate handling and interrogation of these sites if they are to be used. The use of such devices should be cleared with the oncology team, parents, and the surgical team caring for the line. A nasogastric or orogastric tube is usually needed—initially for decompressing the stomach and potentially following surgery for nutrition.

Medication A broad-spectrum antibiotic that covers oral/nasal flora is routine and should be continued in the perioperative period. Other medications to consider for postoperative comfort are antiemetics and pain medications. The surgeon should communicate early with the anesthesiologist about the use of vasopressors. Too often, a wide-open arterial anastomosis has been redone only to find that the agent responsible for the pale flap was the vasopressor. Fluid, colloid, or blood product administration should be the first line of treatment in these cases.

Technical Considerations

Several important technical considerations are related to the actual execution of the operation merit discussion. Careful attention to these issues separates the good outcomes from the potential disaster cases.

Oral Cavity Separation One of the most difficult complications of oropharyngeal reconstruction is the dreaded fistula [3–12]. Fistulas may develop between the oropharynx and the nasal cavity or the skin. Typically, they occur at the flap and native mucosa juncture. To minimize the risk of fistulas, one should consider the causative factors: poor healing and inadequate seal. Poor healing may result from ischemia, infection, or a suboptimal environment (such as bathing in saliva or a radiated tissue bed). Ischemia can be controlled by bringing healthy, well-vascularized tissue to the defect and by resecting all poorly perfused tissues. Inadequate seal is almost always a result of poor surgical planning or execution. The most problematic areas for obtaining a tight seal are at the gingiva, the palate, and posterior mouth. Patients with intraoral tumor involvement, radiation, or poor oral hygiene may present with mucosa that is friable. The right approach is to remove all of the friable and suboptimal tissue from the area so that a tight seal can be created with the newly transferred flap and the surrounding tissues.

Brain–Mucosa Separation When reconstructing defects that involve the cranial base, it is critical to obtain a good seal to separate the brain from the mucosa [13]. Tumor extirpation operations that involve the cranial base typically leave a large soft tissue defect. Obliteration of the resulting dead space is paramount to avoid cerebrospinal fluid leakage and infection. It is not uncommon to have to utilize a muscle flap in addition to a fasciocutaneous flap in these cases—the former for obliteration of the dead space and the latter for mucosal reconstruction.

Microsurgery The importance of adequate vessels for microvascular anastomosis cannot be overstated—the larger the vessels, the higher the likelihood of success. Source

vessels found in the neck have reliable anatomy and flow. These vessels may be too distant for more cephalic defects such as the scalp or orbit; in which case, the facial or superficial temporal vessels may be substituted [14]. In head and neck reconstruction, one rarely encounters difficulty in finding a suitable artery. However, finding an appropriate vein can sometimes be challenging. Good communication between the extirpative team and the reconstructive team from the outset of the procedure may allow for the identification and preservation of useful recipient vessels later in the procedure. In situations where the area is heavily scarred or has been previously radiated, one should consider (a) vein grafting to the opposite side or (b) use of the ipsilateral cephalic vein. It is rarely worth the risk to use less than optimal vessels in a zone compromised by scarring or radiation, to avoid the additional effort of vein grafting, using the contralateral side or the ipsilateral cephalic vein. We have found the cephalic vein quite useful in difficult outflow situations. A long segment can be harvested from the ipsilateral arm using multiple stab incisions. Minimal morbidity, anatomic consistency, and long length make this vein a perfect "bail out" strategy in difficult situations. There is ongoing debate in the literature about immediate versus delayed use of arteriovenous loops. The most recent literature suggests that staging of arteriovenous loops is not necessary [15].

Flaps

In this section, we will outline common flaps that are utilized for head and neck reconstruction. These flaps have consistent anatomy, low donor site morbidity, and long, reliable pedicles that allow a wide reach in the head and neck—they are the workhorse flaps of head and neck reconstruction [16].

Radial Forearm Flap [17, 18] This flap provides thin, reliably perfused tissue based on a long pedicle for reconstruction of small to moderate sized defects. The anatomy is consistent, the flap is easy to harvest, and outcomes have been excellent [6, 7, 11, 12, 16, 19–21]. It can be harvested as a fasciocutaneous flap or an adipofascial flap. Inclusion of the medial or lateral antebrachial cutaneous nerve creates a neurosensory flap that may be useful, to restore sensation to areas such as the palate. For small flaps, the donor site can be closed linearly. For larger flaps, a skin graft is required. The healing of this skin graft can be problematic if the paratenon over the flexor carpi radialis tendon is stripped [21–24]. Prior to harvesting this flap, one must perform an Allen's test to confirm integrity of the superficial palmar arch.

Anterolateral Thigh Flap Based on the descending branch of the lateral femoral circumflex artery, this versatile flap provides a substantial surface area of skin for reconstruction of large defects in the head and neck [16, 25–30]. The anatomy of the flap and pedicle are reliable and consistent. A large amount of skin and subcutaneous fat can be harvested with the flap and the donor site morbidity is minimal [31]. In some cases, the vascular pedicle courses along the fascial interface between the rectus femoris and the vastus lateralis muscles. However, in most cases, the vascular pedicle is intramuscular, thus making the dissection more tedious. In larger patients, its relatively remote location from the head and neck, as well as its anterior location makes it amenable to a two-team approach.

Rectus Abdominis (Myo or Myocutaneous) This flap is used in a variety of anatomic locations and in head and neck reconstruction can provide cutaneous coverage or fill large cavities (Figs. 2.2–2.7) [6, 8, 13, 32]. The flap is harvested from the lower abdomen, preferably through a low transverse incision when only muscle is required or with an ellipse of skin and fat contiguous with the underlying muscle when coverage or lining is needed. The blood supply to the flap is via the inferior epigastric system. The pedicle is typically large, long, and easy to dissect. Depending on the amount of fascia taken with the muscle, the abdominal defect can be prepared directly or with a small mesh patch. Attention must be paid to proper closure as bulges or hernias may result. Abdominal wall function and trunk support is not impacted as long as the contralateral rectus muscle is functional. When placed low enough, the donor site scar is fairly inconspicuous.

Fibula Flap (Osseous or Osseofasciocutaneous) This is another workhorse of head and neck reconstruction, especially in cases where bone is needed (Figs. 2.8–2.10) [3, 6, 16, 33–35]. The fibula flap relies on the peroneal vascular pedicle for blood flow. Dissection of this flap requires thorough anatomical knowledge of the leg and its neurovascular structures—so as to recover a healthy flap and to avoid injury to normal structures. Dissection of the skin flap along with the bone (osseofasciocutaneous flap) can be a bit more cumbersome given that there is a very thin fascia separating these structures, and the number and caliber of perforators within this fascia can be few and small, respectively. However, the long leash of the fascia provides significant versatility in positioning the skin appropriately to fit the given defect.

Summary

Reconstructive surgery is an integral part of treatment for children with head and neck tumors. Inclusion of the reconstructive surgeon at the outset of treatment improves the likelihood of an optimal outcome by facilitating interdisciplinary

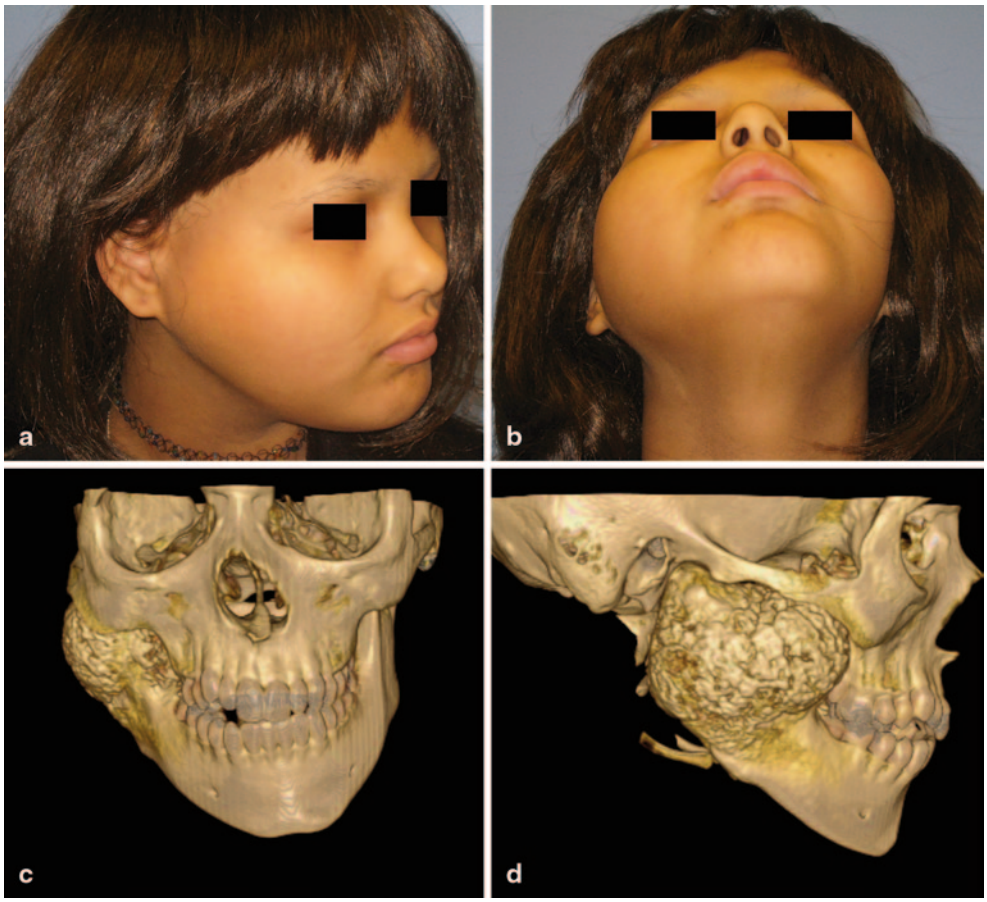

Fig. 2.2 Ten-year-old girl following neoadjuvant chemotherapy and radiation for a high grade osteogenic sarcoma of the right mandibular body (**a, b**). The 3D maxillofacial CT scans (**c, d**) demonstrate the large tumor extending up to and involving the adjacent skull base on the affected side

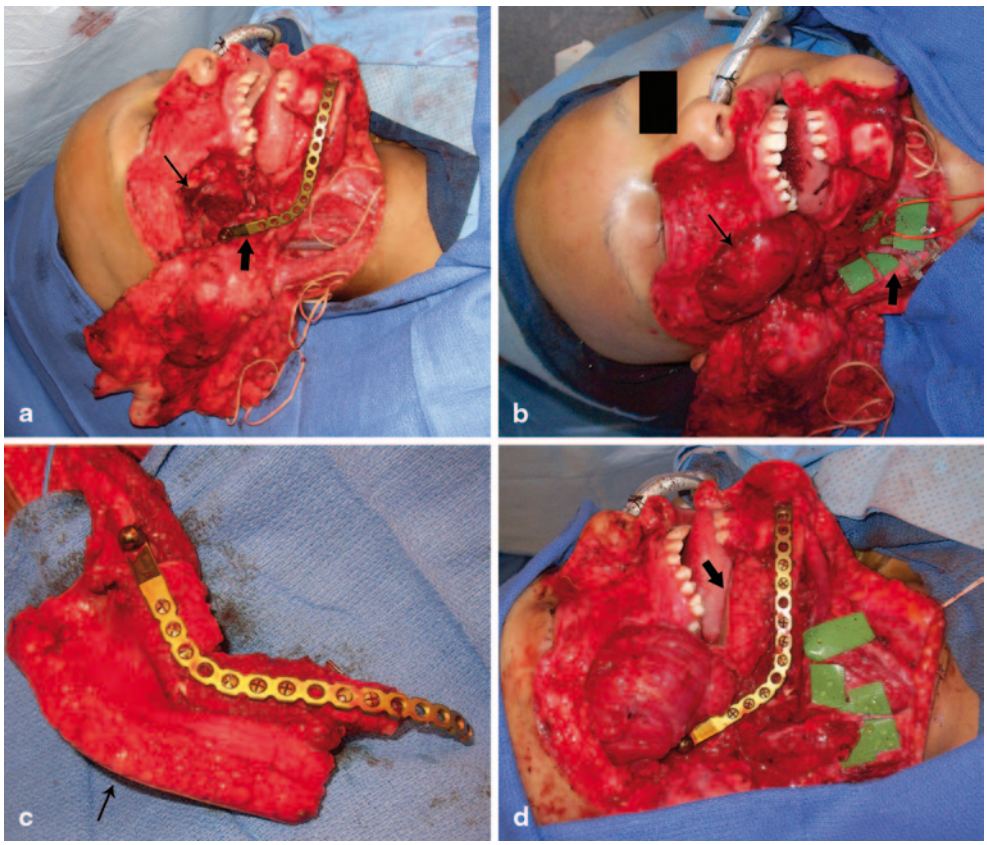

Fig. 2.3 **a** Following extirpation of this tumor via an extended Weber–Ferguson approach, loss of soft tissue and bone has created a large void adjacent to the infratemporal fossa (*thin, black arrow*). A mandibular reconstruction plate has been placed to demonstrate the absent right hemimandible (*thick, black arrow*). **b** A rectus abdominis muscle flap has been inset into the large skull base defect (*thin, black arrow*) and microvascular coaptations have been performed (*thick, black arrow*). **c** An osseofasciocutaneous fibula flap has been contoured and fixed to a mandibular reconstruction plate bent preoperatively to match the contralateral side (*thin, black arrow*). The skin paddle and soft tissue are shown inferiorly (*short, black arrow*). **d** Both flaps have now been inset. The closure of the oral lining has been completed prior to skin closure to allow for a meticulous two-layer closure under direct visualization (*thick, black arrow*)

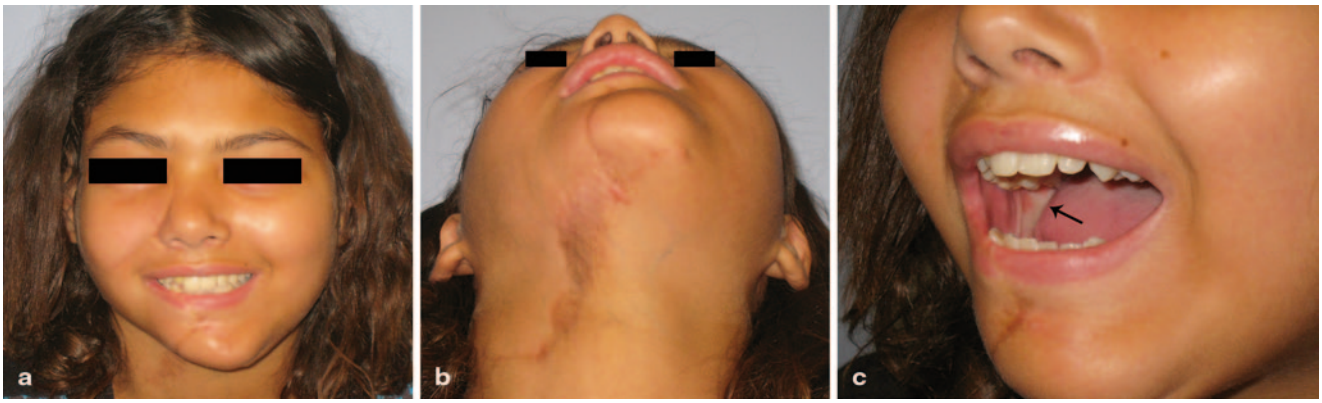

Fig. 2.4 Three years postoperatively. **a** The anteriorposterior (AP) view demonstrates some chin asymmetry secondary to differential right and left mandibular growth and soft-tissue loss on the right side. **b** Submental view demonstrating widened scarring where secondary local tissue rearrangement was required because of native skin flap loss. **c** Some degree of temporomandibular joint (TMJ) stiffness with maximal interincisal opening of 23 mm. The cutaneous portion of the flap (*thin, black arrow*) is well-healed to the adjacent pink oral mucosa

Fig. 2.5 Seventeen-year-old male who presented with swelling on the right side of his face (**a, b**). An axial (**c**) and coronal (**d**) computed tomogram demonstrate an expansile mass obliterating the right maxillary sinus. The 3D CT (**e**) view demonstrates the extent of the lesion and marked thinning of the maxillary bone. A transgingival biopsy revealed an odontogenic myxoma

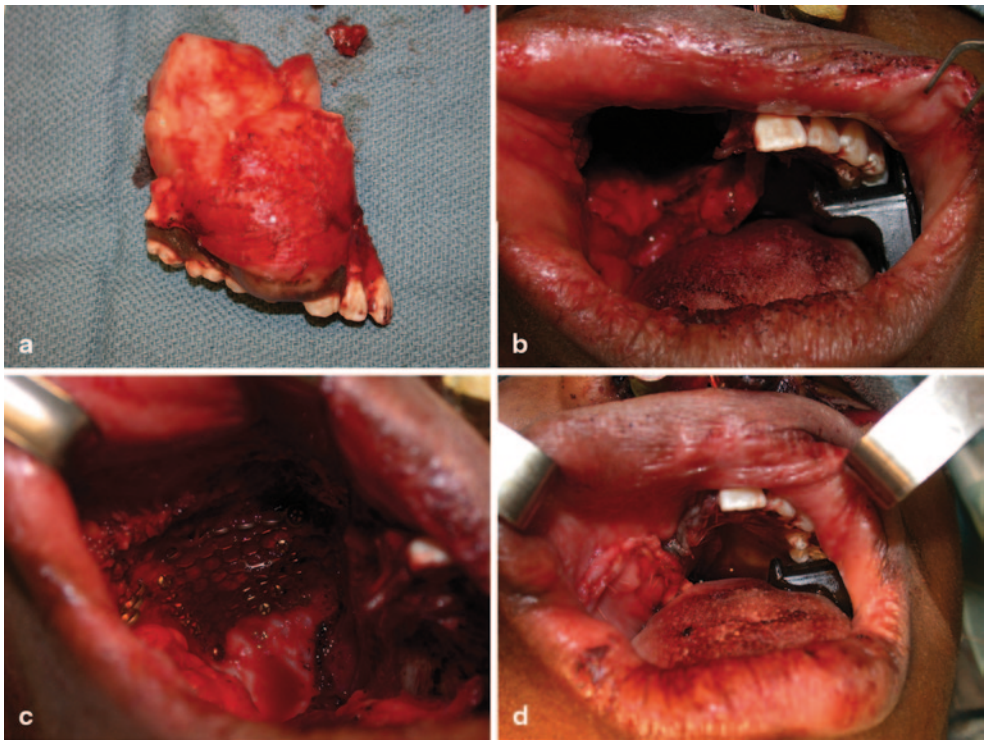

Fig. 2.6 **a** The specimen following an entirely transoral resection. **b** The resultant voluminous defect extending up to and including the orbital floor. **c** Titanium mesh plates have been placed to support the globe and a rectus myocutaneous flap was used to fill the sinus and separate the sinus and oral cavity from hardware (not shown). **d** Following closure

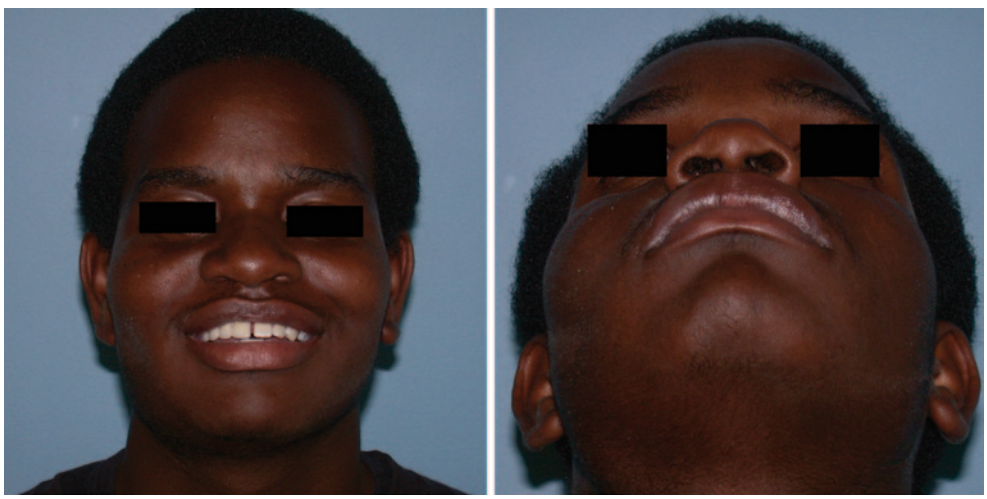

Fig. 2.7 One year after surgery with a partial denture in place (**a**). Some flattening of the right cheek and mild enophthalmos are appreciated on the submental view (**b**) but were not clinically significant

communication and integrating the reconstructive treatment into a long-term care plan. Specifically, the anatomic requirements of the reconstruction can be articulated by the extirpative team, and the rationale for, and timing of adjuvant therapy can be worked out. Preoperative consultation by the reconstructive team provides the opportunity to assess the unique patient factors (e.g., comorbidities, available donor sites, family support) that help determine the most appropriate type of reconstruction. Intraoperative coordination between surgical teams and anesthesia is also vitally important. Patient positioning, type and location of lines and tubes, and simultaneous versus staggered surgery between extirpative and reconstructive teams should be agreed upon. Although many local, regional, and distant flaps exist, a select group of workhouse flaps are commonly used. Special attention should be paid to sealing potentially problematic areas such as the oral cavity, sinuses, or cranium. The reconstructive process in the pediatric patient does not end at discharge but often extends over years. The effects of growth and time often mandate revisions as the child ages and this possibility should be fully disclosed to families at the initial consultation. Although successfully treating the patient's

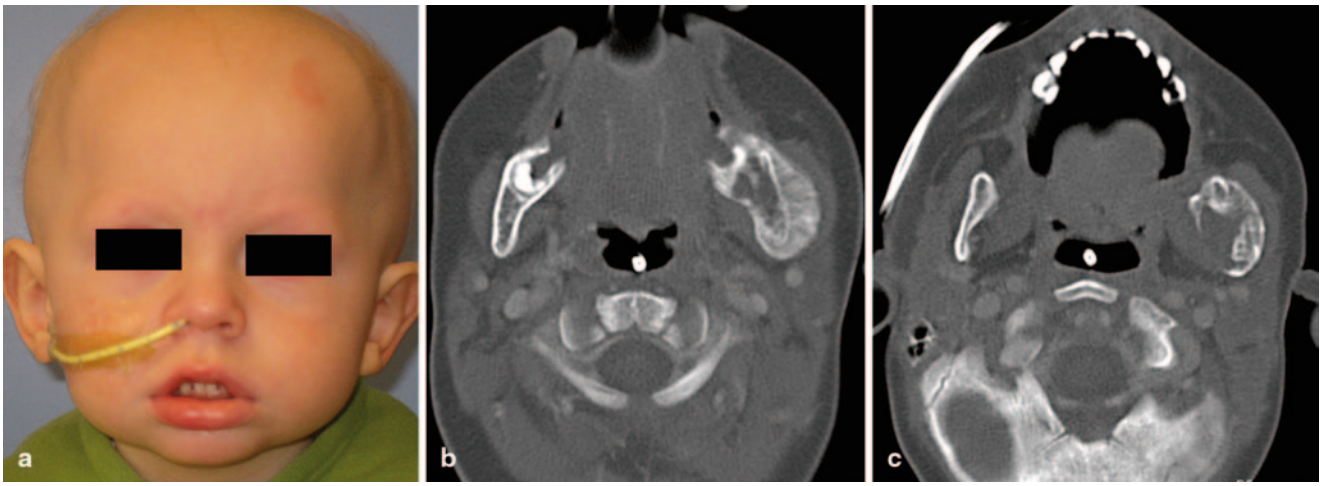

Fig. 2.8 Two-year-old boy after neoadjuvant chemotherapy for a Ewing sarcoma of the left mandible (**a**). Axial computer tomograms of the tumor involving the left mandibular body prior to (**b**) and following chemotherapy (**c**). Because of the proximity of the tumor to the oral lining, it was felt that autogenous reconstruction rather than a temporary reconstruction plate would be required

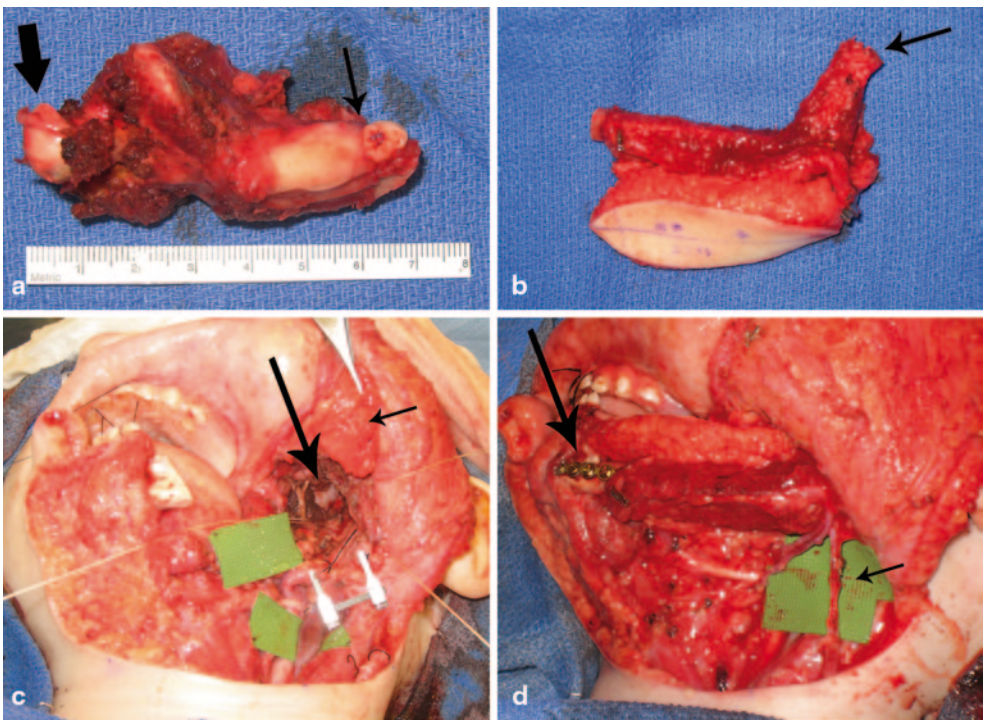

Fig. 2.9 **a** The specimen following extirpation. The condyle (*thick, black arrow*) and the oral lining and dentition (*thin, black arrow*) are seen. **b** Osseofasciocutaneous fibula flap has been harvested and contoured. The new condyle has been contoured and covered with vascularized muscle and periosteum to diminish chances of ankylosis (*black arrow*). The single osteotomy at the angle of the construct was fixed with a resorbable plate to facilitate future distraction (not shown). **c** Reconstruction of the temporomandibular joint was accomplished using vascularized buccal fat pad (*short, black arrow*) and resorbable suspension sutures to hold the new condyle in position. The glenoid fossa seen at the depths of the incision (*long, black arrow*) was not involved. **d** The mandibular construct has been inset with the distal fixation at the left parasymphysis visible (*long, black arrow*). Microvascular anastomoses between the peroneal artery and its two venae comitantes and the facial artery, facial vein, and an external jugular vein are shown (*short, black arrow*)

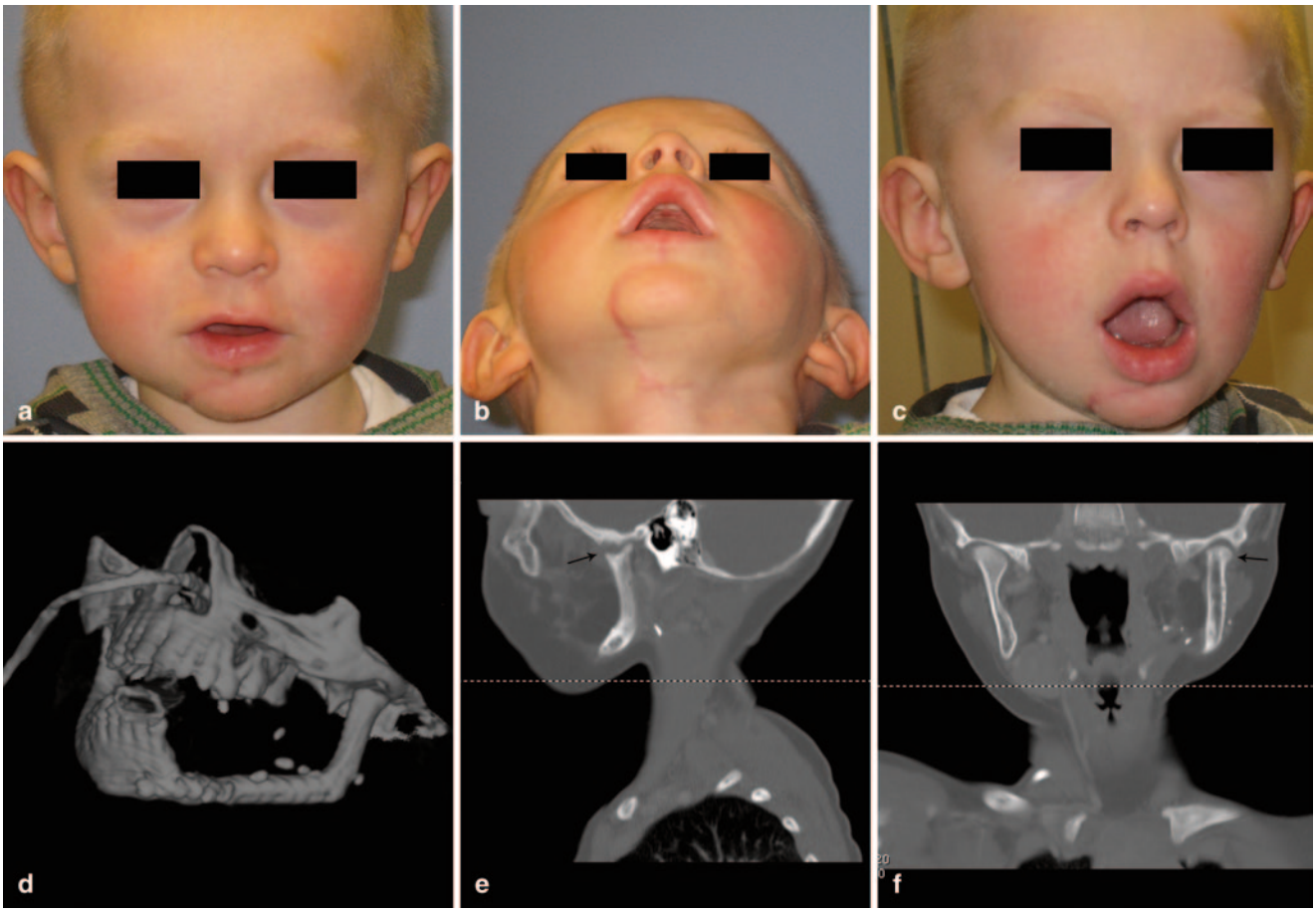

Fig. 2.10 Seven months postoperatively. The AP and submental views demonstrate healing incisions and good symmetry (**a, b**). There were no limitations in mouth opening noted on examination or by the parents (**c**). Postoperative 3D computed tomogram demonstrating the mandibular construct (**d**). Sagittal (**e**) and coronal (**f**) computed tomograms demonstrate the reconstructed condyle well-seated in the glenoid fossa with adequate joint space between condyle and glenoid seen (*long, black arrows*)

tumor remains the primary goal of therapy, the quality of the life that has been saved will be improved by a well-planned and well-executed reconstruction.

References

1. Markowitz BL, Calcaterra TC. Preoperative assessment and surgical planning for patients undergoing immediate composite reconstruction of oromandibular defects. Clin Plast Surg. 1994; Jan;21(1):9–14.
2. Duncan MJ, Manktelow RT, Zuker RM, Rosen IB. Mandibular reconstruction in the radiated patient: the role of osteocutaneous free tissue transfers. Plast Reconstr Surg. 1985 Dec;76(6):829–40.
3. Cordeiro PG, Disa JJ, Hidalgo DA, Hu QY. Reconstruction of the mandible with osseous free flaps: a 10-year experience with 150 consecutive patients. Plast Reconstr Surg. 1999 Oct;104(5):1314–20.
4. Cordeiro PG, Hidalgo DA. Soft tissue coverage of mandibular reconstruction plates. Head Neck. 1994 Mar–Apr;16(2):112–5.
5. David DJ, Tan E, Katsaros J, Sheen R. Mandibular reconstruction with vascularized iliac crest: a 10-year experience. Plast Reconstr Surg. 1988 Nov;82(5):792–803.
6. Hidalgo DA, Disa JJ, Cordeiro PG, Hu QY. A review of 716 consecutive free flaps for oncologic surgical defects: refinement in donor-site selection and technique. Plast Reconstr Surg. 1998 Sep;102(3):722–32 (discussion 733–24).
7. Hidalgo DA, Pusic AL. Free-flap mandibular reconstruction: a 10-year follow-up study. Plast Reconstr Surg. 2002 Aug;110(2):438–49 (discussion 450–31).
8. Kroll SS, Baldwin BJ. Head and neck reconstruction with the rectus abdominis free flap. Clin Plast Surg. 1994 Jan;21(1):97–105.
9. Kroll SS, Goepfert H, Jones M, Guillamondegui O, Schusterman M. Analysis of complications in 168 pectoralis major myocutaneous flaps used for head and neck reconstruction. Ann Plast Surg. 1990 Aug;25(2):93–7.
10. Panje W. Immediate reconstruction of the oral cavity. In: Thawley S, Panje W, editors. Comprehensive management of head and neck tumors. Vol I. Philadelphia: Saunders; 1987. p. 563.
11. Schusterman MA, Miller MJ, Reece GP, Kroll SS, Marchi M, Goepfert H. A single center's experience with 308 free flaps for repair of head and neck cancer defects. Plast Reconstr Surg. 1994 Mar;93(3):472–8 (discussion 479–80).
12. Urken ML, Weinberg H, Buchbinder D, et al. Microvascular free flaps in head and neck reconstruction. Report of 200 cases and review of complications. Arch Otolaryngol Head Neck Surg. 1994 Jun;120(6):633–40.