

March 13, 2016

M.D.

Date of Visit: February 24, 2016

Dear Dr.

We recently had the opportunity to see [REDACTED] when he returned for follow-up consultation accompanied by his mother, [REDACTED] and his sister. Concerns brought to this visit included continuing with his cleft-related care and a stitch in his palate observed by his dentist.

Members of our interdisciplinary team at this visit included:

★ Dr. [REDACTED] Prosthodontist / Program Director
 ★ [REDACTED] Program Coordinator / Speech-Language Pathologist
 Nurse Coordinator
 Dr. [REDACTED] Pediatric Nurse Practitioner
 ★ Dr. [REDACTED] Plastic and Reconstructive Surgeon
 Dr. [REDACTED] and Maxillofacial Surgeon
 Dr. [REDACTED] Otolaryngologist
 Dr. [REDACTED] / Pediatric Dentist – Genetics
 Dental Hygienist
 Dr. [REDACTED] / Resident Pediatric Dentistry
 ★ Dr. [REDACTED] Orthodontist
 Dr. [REDACTED] Resident Orthodontics
 Dr. [REDACTED] / Audiologist
 [REDACTED] Speech-Language Pathologist
 [REDACTED] / Clinical Fellow Speech-Language Pathologist

[REDACTED]'s providers include:

Dr. [REDACTED] / Primary Care Physician
 Dr. [REDACTED] / Plastic and Reconstructive Surgery
 Dr. [REDACTED] / Pediatric Geneticist
 Dr. [REDACTED] / Oral and Maxillofacial Surgeon
 Dr. [REDACTED] / Orthodontist
 Dr. [REDACTED] / Dentist
 [REDACTED] Speech-Language Pathologist

HISTORY

Referral: [REDACTED] was originally seen in this Clinic at the request of Dr. [REDACTED] plastic and reconstructive surgeon, for interdisciplinary evaluation and treatment planning. His last visit to this Clinic was on 2-25-2015.

If not seeking approval as a Craniofacial Team, skip to Standard 2.

Craniofacial (surgery involving a transcranial procedure) Teams must meet Standards 1.1 through 1.3 related to Team Composition, as well as the following Standard.

To view examples of standard 1.4 documentation that Teams have submitted as evidence of compliance of the Standards for Cleft Palate and Craniofacial Teams, please visit: <http://www.acpa-cpf.org/standards-examples>

1.4 The Craniofacial Team must include a craniofacial surgeon who has specialized training and experience in surgical management of patients with syndromic and other craniofacial anomalies. Such training includes surgical procedures for syndromic patients involving the maxillofacial and craniofacial structures, and must include transcranial surgery. In addition, the Craniofacial Team must have access to a psychologist who does neurodevelopmental and cognitive assessment. The results of the neurodevelopmental and cognitive assessment must be part of the CFT team assessment record. The Team also must demonstrate access to refer to a neurosurgeon, ophthalmologist, radiologist, and geneticist. The participation of these individuals should be documented in each patient's team report.

8. List the name of the Team's lead member trained in transcranial craniofacial surgery.
MD, FRCS(C) / Plastic Surgeon

9. Attach a **team report** concerning a patient with a craniofacial condition (requiring transcranial surgery) that documents the participation of the transcranial craniofacial surgeon as well as other team members. **Omit all patient identifying information. Label as 1.4.9** and limit the attachment to five pages or fewer.

10. Describe the process used to obtain evaluation or treatment services by a neurosurgeon, ophthalmologist, radiologist, or geneticist.

Neurosurgery: Dr. _____ and _____ CPNP, complete evaluations in Craniofacial Clinic with craniofacial plastic surgeon. Evaluations are also done more urgently on an inpatient basis and in the neurosurgical ambulatory clinic.
Ophthalmology: Drs. _____ and _____ see patients in _____ with slots held for same-day appointments as needed.
Radiology: Dr. _____ oral and maxillofacial radiologist, provides interpretation of all cone-volumetric CT scans performed for our patients. Other imaging and interpretive services are provided as needed by the _____
Genetics: Dr. _____ medical geneticist, is primary consultant referrals made to and sees patients in the _____ ambulatory clinics.

11. Provide an example of a patient that requires transcranial surgery that documents a referral from the team to a specialist listed in 1.4.10. **Omit all patient identifying information. Label as 1.4.11** and limit the attachment to five pages or fewer.

12. Provide the report from the specialist resulting from the referral. (If you choose imaging, a test alone does not meet the standard. Please demonstrate the interaction/participation of the radiologist.) **Omit all patient identifying information. Label as 1.4.12** and limit the attachment to five pages or fewer.

June 5, 2015

M.D.

Re:

Dear Dr.

Thank you for the opportunity to meet [REDACTED] who was accompanied by his parents, [REDACTED] for a recent consultation regarding concerns of sagittal craniosynostosis.

Members of our interdisciplinary team at this visit included:

Program Coordinator / Speech-Language Pathologist
 Pediatric Nurse Practitioner
 Nurse Coordinator
 ' Craniofacial and Plastic and Reconstructive Surgeon
 ' Neurosurgery
 ' Otolaryngologist
 ' Fellow Speech-Language Pathologist



HISTORY

Referral: [REDACTED] was referred to this Clinic for consultation, including interdisciplinary evaluation and treatment planning by Dr. [REDACTED] pediatric neurosurgeon, with concerns related to his diagnosis of sagittal synostosis. This was his first visit to our clinic.

Diagnosis: Sagittal synostosis (ICD9 756.0)

Social and Family History: [REDACTED] is the second child of his parents; his older sibling is 18 months older and healthy. Family history is reportedly negative for craniosynostosis. A genetics evaluation has not been completed.

Medical and Developmental History: [REDACTED] was born at 31-32 weeks gestation with a birth weight of 3 pounds 15 ounces, length was 42.5 centimeters and his HC was 12.01 inches (30.5 cm). Labor and delivery were complicated by PROM; started on magnesium sulfate for preterm labor. There is a history of polyhydramnios prior to PROM (AFI 27). He was delivered via complicated emergent C-section due to placental abruption. APGAR scores were 3, 2, 6 at 1, 5, 10 minutes, respectively. He was intubated in the OR and received chest compressions for 45xseconds at birth. Emergency vascular access was needed for volume expansion. He was admitted to [REDACTED] at birth. [REDACTED] was discharged to home 55 days after birth (weight 7 pounds 5.4 ounces, height 20.47 inches, HC 35 cm (13.78 inches)).

HISTORY (Continued)

Problem list at discharge was as follows:

- Prematurity at 31-32 weeks
- Respiratory distress syndrome of newborn
- Single suture craniosynostosis: head ultrasound 1/18/15 and skull x-rays 2/23/15
- Atrial septal defect vs PDA ECHO 1/28: no left-right shunt. Murmur on exam. Follow-up planned at 3-6 months
- Stridor & oropharyngeal dysphagia: History of apnea, bradycardia, desaturations, and stridor with feeding. Swallow study 2/26/15 with penetration (no aspiration). NG-tube feedings until 3/7/15. ENT evaluation and OT consulted. No laryngomalacia. Laryngeal edema present. Treated with reflux precautions and 5 day course of Prednisone. Half nectar thick feedings recommend with Dr. Brown slow flow nipple.
- Unspecified nutritional deficiency: poor weight gain. Required fortified feeds Neosure 24k/cal breastmilk
- Observation and evaluation of newborn sepsis (resolved)
- Hypotension: placenta abruption. Currently normotensive
- Neonatal hypoglycemia: Initial blood sugar 28
- Anemia of prematurity: Nadir CT 26 on 2/14. Post blood transfusion 2/15. EPO and folate 2/24-3/16

Surgical History: None to date, status-post blood transfusion 2/26/2015

Medications: Prilosec bid

Allergies: NKDA

RECORDS OBTAINED

Imaging: 3D Head CT scan

Photographs: Facial; profile

Speech: Clinical Feeding Evaluation & Receptive-Expressive Emergent Language Test, 3rd Edition

Hearing: Distortion product otoacoustic emissions

FINDINGS

NURSING / PEDIATRICS

██████████ was seen today with his parents, who describe him as a “happy, smiley, normal baby,” despite a complicated early medical history as described above. He has an abnormal head shape and parents are interested in knowing what the plan will be for managing this. He has a history of heart murmur and is followed in cardiology, and his next appointment is in 2 weeks. Review of systems today is significant for mild congestion following feeds. He is taking Similac Total Comfort, up to 4 ounces per feed over 20 minutes. He is not having projectile vomiting. He is sleeping well without breathing concerns. No cyanosis has been observed. He passed his newborn hearing screen and parents feel he hears them and tracks visually. They describe him as “strong,” and he has been rolling from front to back. He has good head control. Cranial index is 67%. Development is age appropriate based on an adjusted age of 6 weeks in all domains based on the Ireton Child Development Review. He is receiving early intervention services, including occupational therapy. He is followed by Dr. ██████████ and was last seen for his 3-month well child check. He is scheduled for his 4-month check on June 5. He is reportedly up-to-date on immunizations.

SPEECH-LANGUAGE PATHOLOGY

██████████ is a 3 month, 13 day male who has a corrected age of 1 month, 13 days. He was seen for a speech-language evaluation and feeding consultation as a part of his craniofacial team visit today. He has a complicated feeding history and requires a pre-operative language assessment prior to surgical management of surgical correction of his sagittal craniosynostosis. Medical and family history is as described above.

SPEECH-LANGUAGE PATHOLOGY (Continued)

Receptive-Expressive Emergent Language Test, 3rd Edition: The REEL-III provides an estimate, based on parent report, of emergent receptive and expressive language skills and can be used to identify infants and toddlers with major communication problems. It is normed on children from birth to 36 months of age. This was completed by [REDACTED]'s mother and guided by the clinician. Corrected age of 1 month, 13 days was used due to prematurity.

	<u>Raw Score</u>	<u>Ability Score</u>	<u>Percentile Rank</u>	<u>Descriptive Rating</u>
Receptive Language:	7	82	12	Below Average
Expressive Language:	6	89	23	Below Average
Total Language:	-	83	13	Below Average

Receptive Language: [REDACTED] has several receptive language skills emerging in the 0 to 6 month range, such as looking and smiling at those who are talking and localizing toward new sounds. He is not yet shifting eyes between speakers, looking at the speaker's mouth instead of their whole face, or looking for a speaker that is out of sight.

Expressive Language: [REDACTED]'s production of sounds includes long 'E' and short 'a' vowels but has not yet begun to include other sounds, such as "ooo" or any consonant sounds. He has several expressive language skills emerging in the 0 to 6 month range, such as making different sounds for happy versus upset. He is not yet vocalizing back when he hears voices, laughing when others play with him, or making more sounds when he is face-to-face with people, as opposed to being alone.

This assessment reveals mild delays in both receptive and expressive language. This will require monitoring. We recommend audiological assessment and follow-up speech-language assessment in this Clinic in 6 months.

Feeding: [REDACTED] was recently hospitalized with H. influenza. Mother reports his chest x-ray was not normal. Mother reports he was changed from nectar thick feeds to thin based on clinical presentation, without repeat VFSS. Orofacial exam revealed mild ankyloglossia. No signs of a submucous cleft palate. On room air without distress, mild upper airway noise, possible stridor prior to feeding. Fed with Dr. Brown slow flow, thin liquid in upright position. Intake was okay with 10mL in 2 minutes but possible fatigue. Anterior loss of liquid with disorganized SSB rhythm. Cough x 1 and wet. Upper airway congestion/stridor. No nasal regurgitation. Disorganized SSB rhythm.

Impression of possible penetration or aspiration based on cough 1x, vocal wetness and increased upper airway noisiness / congestion. Recommend follow-up with pediatric ENT and speech-language pathology for feeding therapy and upper airway assessment. I am concerned he needs his formula thickened. He may benefit from positioning techniques and a different bottle as well. Consideration may need to be given to additional imaging. I am happy to see him back on July 23rd for follow-up if there are persisting concerns.

AUDIOLOGY

[REDACTED]'s parents reported that he passed his newborn hearing screening. Today, he passed a distortion product otoacoustic emission screening in the right ear and in the left ear except at 5000 HA. He was making quite a bit of noise during testing, which may have compromised the results. [REDACTED]'s hearing and middle ear status should be monitored every 4-6 months until he is 5 years of age.

ENT

[REDACTED] has a history of GERD and stridor with dysphagia. He has been treated with GERD precautions and is currently on Prilosec. Previous laryngeal exam has shown laryngeal edema, treated with a 5-day course of Prednisone in March. Exam today showed normal external ears. Tympanic membranes are intact and dry. Anterior nasal air way is patent. Oral cavity and pharynx appear within normal limits. Neck is supple and without lymphadenopathy. [REDACTED] can follow-up locally for his feeding and continued management of his upper airway/GERD.



CRANIOFACIAL SURGERY

██████ has a complicated medical history as described above. He was hospitalized again for the H. influenza last month. Exam today shows an infant with significant scaphocephaly with sagittal synostosis. This was confirmed by a head CT scan today. Cranial measurements are: 92 mm transverse and 144 anterior-posterior with cranial index of 0.64. Bilateral craniotomies are recommended in late July or August and will be coordinated with Dr. ██████ and me. The risks associated with this procedure were discussed, as well as the perioperative and post-operative course. Risks include bleeding, transfusion, infection, and need for revision surgery. There are cases of death reported in the literature as well. I answered all parents' questions today to the best to my ability. My office will contact ██████'s parents to arrange surgery. They can contact us if they have any additional questions prior to surgery.

NEUROSURGERY

██████ was born 8 weeks premature. He has a complicated medical history as described above. He was recently hospitalized again for the H. influenza, discharged 4/13. He is eating well every 3 to 4 hours per mother, but is noisy. He has reflux but no vomiting. He is sleeping well. He wakes on his own to eat. Lethargy is denied by his mother. He had a head CT today confirming sagittal craniosynostosis. Physical exam shows an alert and smiley infant. He has frontal bossing with symmetric facies. Ears are well aligned. Extraocular movements are intact, and movement of extremities is normal. Cranial measurements include: 141 mm AP, 94 mm transverse, 131 mm right oblique, 135 left oblique. Cranial index is 0.67. We discussed surgical repair of his sagittal synostosis with Drs. ██████ and ██████ sometime this summer.

CARE PLAN

1. ██████ should continue follow-up monitoring of growth and development with his primary care physician and his specialists, including cardiology for his ASD. He needs close follow-up of problems related to his prematurity.
2. ██████ is scheduled for bilateral craniotomies to repair his sagittal craniosynostosis with Dr. ██████ craniofacial plastic surgeon, and Dr. ██████ neurosurgeon, on August 18, 2015 at the ██████. Dr. ██████'s nurse will contact ██████'s parents with pre-operative instructions.
3. Prior to surgery, ██████ will need a complete pre-operative ophthalmological examination. ██████ should be coordinated locally if possible. Results can be faxed to the attention of Neurosurgery at ██████.
4. ██████ needs follow-up pediatric ENT and feeding assessment for oropharyngeal dysphagia and stridor in the near future. He should continue treatment of his GERD. Consideration may need to be given to thickened feeds, positioning, and repeat imaging. He can be seen for follow-up in this Clinic with ██████ on July 23, 2015 if there are persisting concerns (██████).
5. A genetics evaluation is recommended for ██████. This is scheduled for Thursday ██████ 15 at 12:45 p.m. with Dr. ██████ at the ██████.
6. ██████ should continue receiving Early Childhood Special Education Services.

We will plan to see ██████ for a post-operative visit on September 1, 2015 at 2:00 p.m. Thank you for allowing us to share in his care. Please do not hesitate to contact us with any questions or concerns (██████-██████).

Program Coordinator
cc: Dr. ██████
Copy Via ██████

CARE PLAN

1. █ should continue regular well-child checks. It is important growth and development be monitored closely. A referral to Birth-to-3 has been provided. The family has also been given provided information to schedule follow-up with the Genetics Clinic.
2. █ is scheduled for craniotomy surgery with Dr. █ and Dr. █ on April 10, 2017. A surgical nurse will call within a week prior to surgery to discuss in more detail.
- ★ 3. █ is scheduled to see Dr. █ pediatric ophthalmologist in the █ on Wednesday, March 8th, 2017 at 10:20 a.m. It is important this evaluation is completed prior to craniovault remodeling surgery.
4. █ should continue follow-up audiology and ENT care. This could be arranged with Dr. █ (█).

We will see █ back in our clinic approximately one month post-surgery. Thank you for allowing us to share in her care. Please do not hesitate to contact us with any questions or concerns (█).

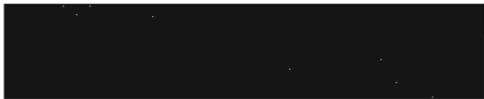
Interim Program Director

Program Coordinator

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- Dr. █ / Pediatrician
- Dr. █ / Craniofacial and Plastic and Reconstructive Surgeon
- Dr. █ / Neurosurgeon
- Dr. █ / Otolaryngologist
- Dr. █ / Pediatric Ophthalmologist

Parents:



Attachment: Audiogram

Office Visit 3/8/2017

Provider: MD (Ophthalmology)

Primary diagnosis: Obstruction, nasolacrimal duct, neonatal, left

Reason for visit: Crouzon's Disease; Referred by

MD

Progress Notes

MD (Physician) • Ophthalmology

Chief Complaints and History of Present Illnesses

Patient presents with

• **Crouzon's Disease**

Patient is to have surgery to fix synostosis on April 10. Here to have left eye evaluated to determine if there is anything that needs to be done with it. Mom has noted patient frequently rubbing left eye since birth. Also notes discharge for past 2-3 months. No massaging to date, no drops. No strab noted. Previously had left face turn, but this has resolved per mom. Mom and sister have more severe Crouzon.

CT HEAD W/O CONTRAST 9/30/2016 1:28 PM

History: eval for synostosis, Congenital malformation of skull and face bones, unspecified

Comparison: No prior similar studies.

Technique: Using multidetector thin collimation helical acquisition technique, axial, coronal and sagittal CT images from the skull base to the vertex were obtained without intravenous contrast.

Findings: Motion artifact degrades image quality.

No intracranial hemorrhage, mass effect, or midline shift. The ventricles are proportionate to the cerebral sulci. The gray to white matter differentiation of the cerebral hemispheres is preserved. The basal cisterns are patent. There is near complete closure of the a left coronal suture with resultant anterior plagiocephaly and scalloping of the cranial vault right greater than left.

The visualized paranasal sinuses are clear. The mastoid air cells are clear.

Impression: Unilateral left coronal synostosis.

Review of systems for the eyes was negative other than the pertinent positives and negatives noted in the HPI.
History is obtained from the patient and mom.

: No primary care provider on file.

is home

Assessment & Plan**Assessment & Plan**

old female who presents with:

Crouzon syndrome

asymmetric with LEFT unilateral frontal craniosynostosis

set to have surgery 4/10/16 with Dr.

Enophthalmos and congenital ptosis, LE

ptosis of upper lid associated with enophthalmos, but doesn't block visual axis
- monitor for now, consider oculoplastics in future

Esotropia, Strabismic amblyopia, LE

- begin patching right eye 2 hours daily

Hypermetropia, bilateral

mild, observe

NLDO, congenital, Left

Exam consistent with total obstruction, may have bony hypoplasia on L.

- massage, polytrim as needed, probing needed - attempt to combine with Dr. s procedure

- I recommend bilateral probing & irrigation with possible stent placement and inferior turbinate in-fracture. Today with her Mom, I reviewed the indications, risks, benefits, and alternatives of bilateral probing & irrigation of the nasolacrimal systems with possible stent placement and possible inferior turbinate in-fracture including, but not limited to, failure to resolve tearing and need for additional surgery, creation of a false passage, and changes in eyelid position. We also discussed the risks of surgical injury, bleeding, and infection which may necessitate further medical or surgical treatment and which may result in diplopia, loss of vision, blindness, or loss of the eye(s) in less than 1% of cases and the remote possibility of permanent damage to any organ system or death with the use of general anesthesia. I explained that we would hide visible scars as much as possible in natural creases but that every patient heals and pigments differently resulting in a variable degree of scarring to the eyes or surrounding facial structures after surgery. I provided multiple opportunities for questions, answered all questions to the best of my ability, and confirmed that my answers and my discussion were understood.

Return for surgery.

Patient Instructions

Patch the RIGHT eye 2 hours while awake EVERY day.

PATCH THERAPY FOR AMBLYOPIA

Your child is being treated for a condition called amblyopia (visual developmental delay). In nonmedical terms, this is sometimes referred to as "lazy eye." Proper motivation and compliance with the patching schedule is of great importance to the success of the treatment. The following are commonly asked questions about patching.

What type of patch should be used?

We recommend the Opticlude, Coverlet, or Ortopad brands of patches. These fit securely on the face and prevent light from entering the patched eye, as well as reducing the likelihood of peeking over or around the patch. Your pharmacist may order these patches if they are not in stock. They come in junior size for infants and regular size for older children. A patch should not be used more than once. They are usually packaged in boxes of 20. You can make your own patch with a gauze pad and tape, but this is a bit more time consuming and not quite as attractive. The black eye patch that ties around the head is not recommended since it may be easily displaced, and the child may peek around the patch.

When should the patch be applied?