Craniosynostosis v4.0: Diagnostic Phase

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

DIAGNOSTIC PHASE

Guidelines for surgical procedure and timeframe

- Posterior vault distraction osteogenesis (PVDO), early infancy
- Fronto-orbital advancement, 9-12 months of age
- Posterior vault switch cranioplasty, 6-9 months of age
- Fronto-orbital advancement 9-15 months of age
- Modified pi, 4-6 mos
- Posterior cranial vault expansion
- Endoscopic strip craniectomy ≤ 4 months of age

Inclusion Criteria

- Child with Craniosynostosis AND
- Candidate for Cranial Vault Expansion

Exclusion Criteria

- Children with diagnosis other than Craniosynostosis or without Cranial Vault Expansion

Initial Assessment

- Ensure patient meets inclusion criteria AND surgery is recommended
- Determine surgical procedure
- Submit surgical planning sheet
- If PVDO, then also schedule hardware removal date 8-10 weeks post-op

No → **Off Pathway**

Yes ↓

Evaluate
Murmur?
Bleeding disorder?
MRSA?

Yes → **Further assessments**

- If murmur, ECHO and consider cardiology consult
- If personal or family history of bleeding disorder, obtain PT, PTT, INR, Von Willebrand antigen, stored specimen and hematology consultation
- If positive MRSA history, obtain MRSA screening swab

No ↓

Ensure appropriate evaluations have been initiated

- Complete Phase 1 of CIS PowerPlan and Checklist 1

↓

To Pre-Surgical Phase

!

Atrial septal defects should be treated prior to surgery given the high risk for venous air embolism with cranial vault expansio. *PFOs are common and do not require treatment

!

Obtain a cervical CT in patients with syndromic synostosis given the higher risk of cervical anomalies

Last Updated: March 2019
Next Expected Review: March 2024

Craniosynostosis v4.0: Pre-Surgical Phase

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

PRE-SURGICAL PHASE

Guidelines for surgical procedure and timeframe

- Posterior vault distraction osteogenesis (PVDO), early infancy
- Fronto-orbital advancement, 9-12 months of age
- Posterior vault switch cranioplasty, 6-9 months of age
- Fronto-orbital advancement 9-15 months of age
- Modified pi, 4-6 mos
- Posterior cranial vault expansion
- Endoscopic strip craniectomy ≤ 4 months of age

Inclusion Criteria

- Child with Craniosynostosis AND
- Candidate for Cranial Vault Expansion

Exclusion Criteria

- Children with diagnosis other than Craniosynostosis or without Cranial Vault Expansion

Pre-surgical Assessment

- Ensure patient meets inclusion criteria AND surgery is recommended
- Confirm planned surgical procedure

No → **Off Pathway**

Yes

All patients must complete an evaluation in PASS clinic

!

Patients with a bleeding disorder must be cleared by Hematology; Review PASS assessment; Review MRSA screening

Ensure appropriate evaluations have been completed

- Order blood
- Obtain consents:
  - Blood consent
  - Neurosurgery consent
  - Plastic Surgery consent
- Complete Phase 2 of CIS PowerPlan and Checklist 2

Pre-operative teaching and prepare for OR → **To Holding Area Phase**

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Craniosynostosis v4.0: Holding Area Phase

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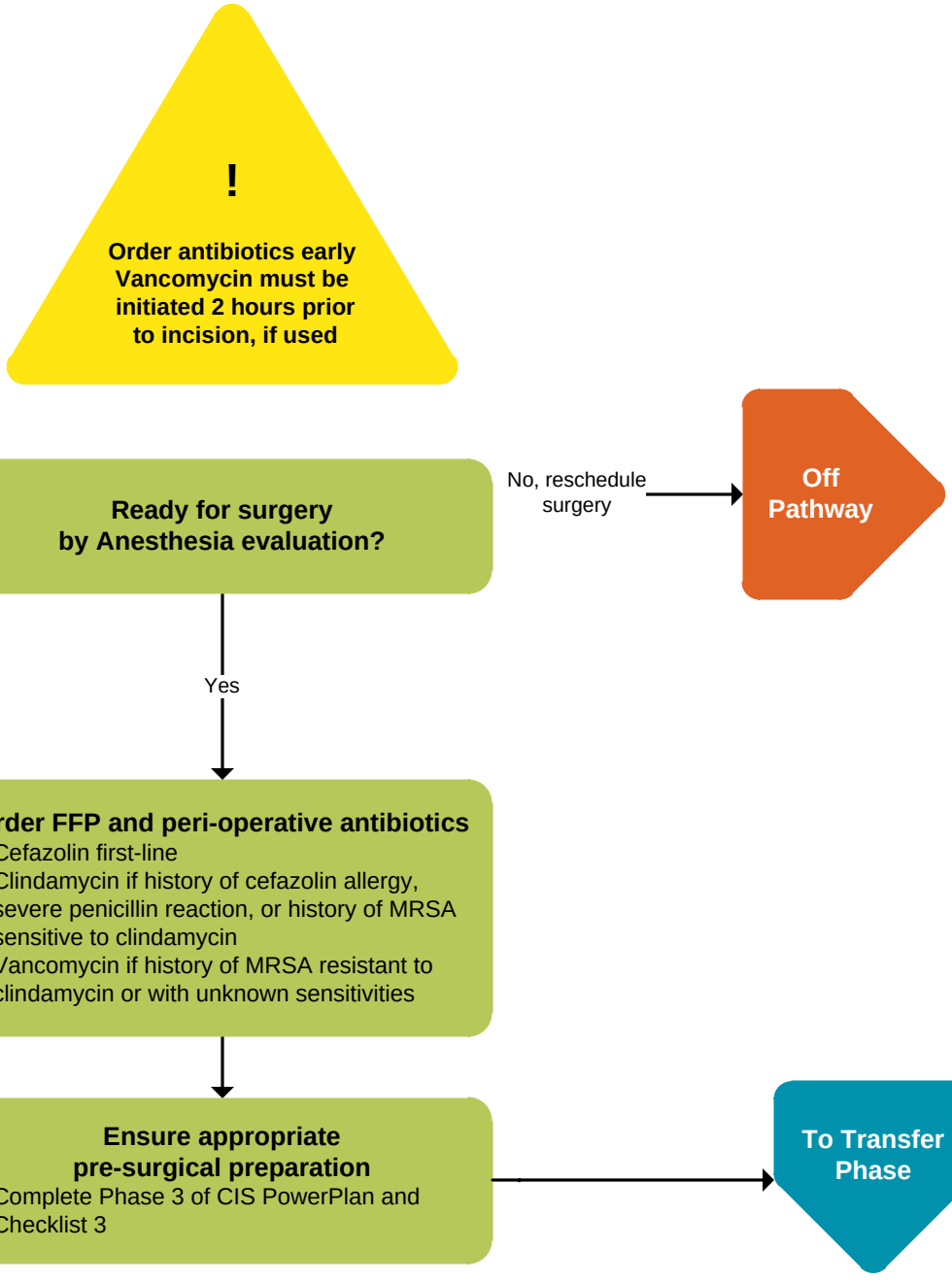
HOLDING AREA PHASE



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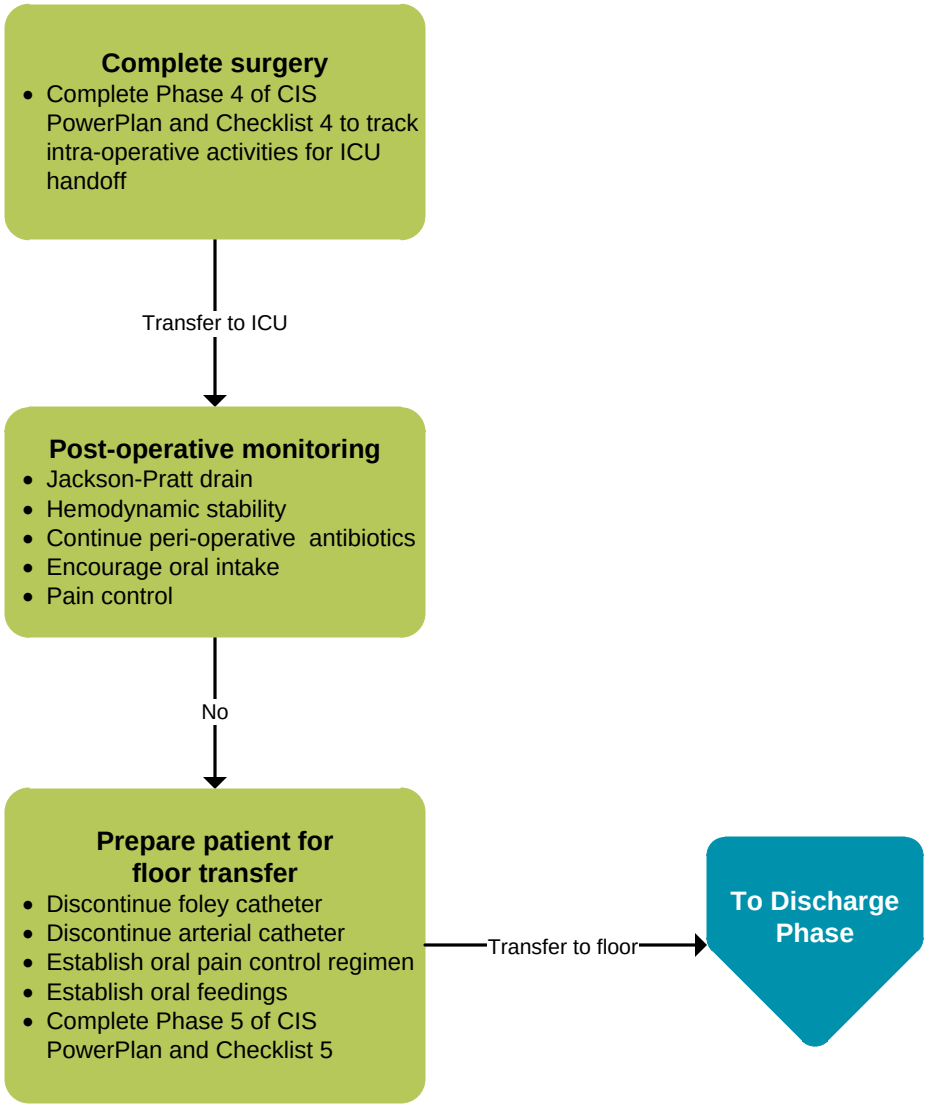
Craniosynostosis v4.0: Transfer Phase

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

PICU TRANSFER AND SURGICAL UNIT TRANSFER PHASES

Continue peri-operative antibiotics

- Cefazolin first-line
- Clindamycin if history of cefazolin allergy, severe penicillin reaction, or history of MRSA sensitive to clindamycin
- Vancomycin if history of MRSA resistant to clindamycin or with unknown sensitivities



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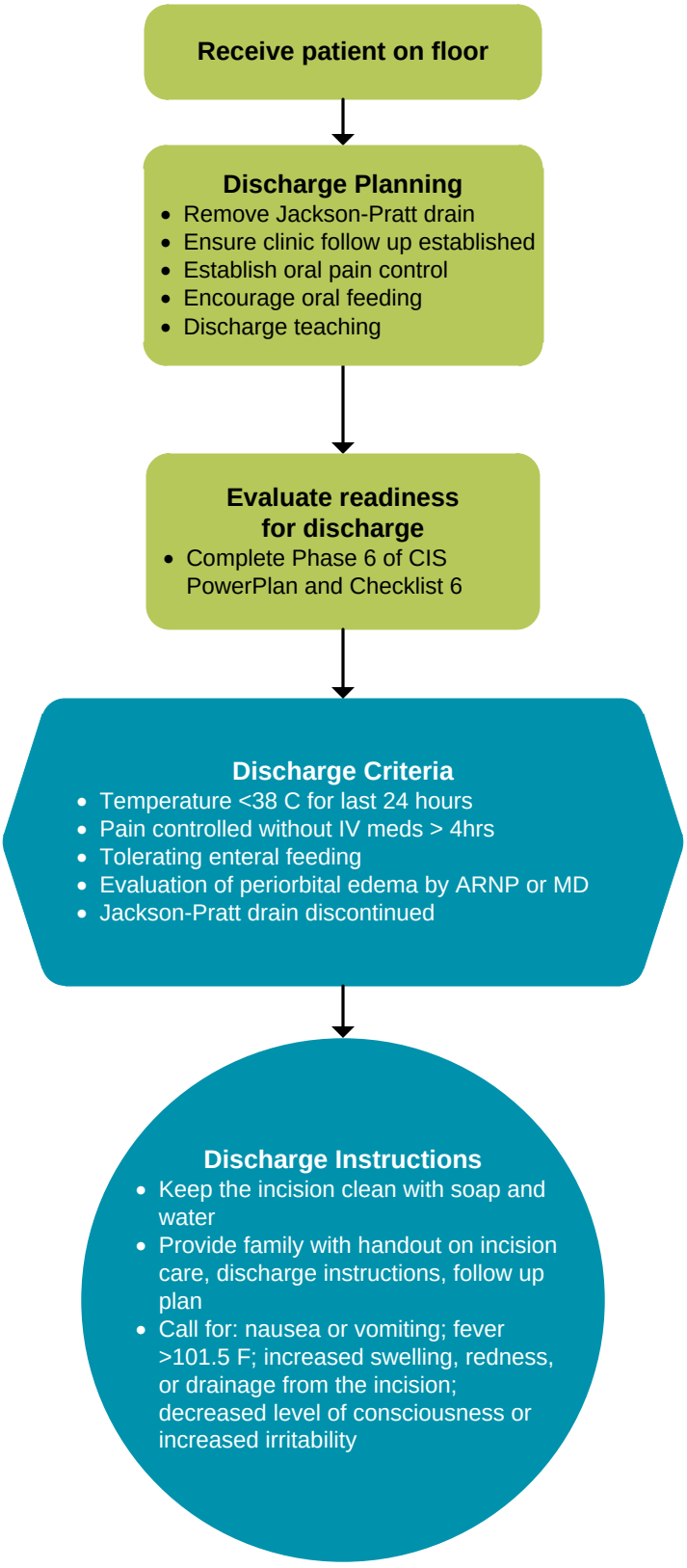
Craniosynostosis v4.0: Discharge Phase

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

DISCHARGE PHASE



Last Updated: March 2019
Next Expected Review: March 2024

Craniosynostosis v4.0: First Year Follow-up

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

Follow-up For the First Year after Cranial Vault Reconstruction Based on Type of Intervention

Inclusion Criteria
- Child with Craniosynostosis AND
- Candidate for Cranial Vault Expansion

Exclusion Criteria
- Children with diagnosis other than Craniosynostosis or without Cranial Vault Expansion

Types of Interventions
- Single Suture Synostosis
- Coronal
- Crouzon/Pfeiffer
- Apert

Initial post-op visits with PS

Fronto-Orbital Advancement
Posterior switch cranioplasty
Modified Pi and Posterior Vault

2-4 weeks post-op
Incision exam

Distraction Osteogenesis of the cranial vault

1 week after hardware placement
- Incision check
- Lateral skull film prior to clinic visit on same day

End of activation phase
(typically 12-16 days)
- Lateral skull film prior to clinic visit on same day
- Removal of activation arms

Operative
- Order CT
- Hardware removal
- Immediate post-op CT
- Same day discharge

2-4 weeks post-op
- Incision exam

Endoscopic

14-21 days (once helmet has been fit)
- Incision check
- Assess helmet fit

ARNP check in with Hanger

Evaluate at end of 1st helmet
- Incision check
- Assess helmet fit

ARNP check in with Hanger

Evaluate at end of 2nd helmet, 3rd needed?
- Incision check
- Assess for 3rd helmet?

Additional surgery?

YES → **Follow path for surgical procedure above**

NO

1 year follow-up with full team
- Shape assessment – 3dMD image
- Developmental assessment
- Clinical assessment for large, palpable skull defects
- Screen: hearing, OSA, vision, concerns for pain, medical and dental home

Single Suture Synostosis | Coronal | Crouzon/ Pfeiffer | Apert

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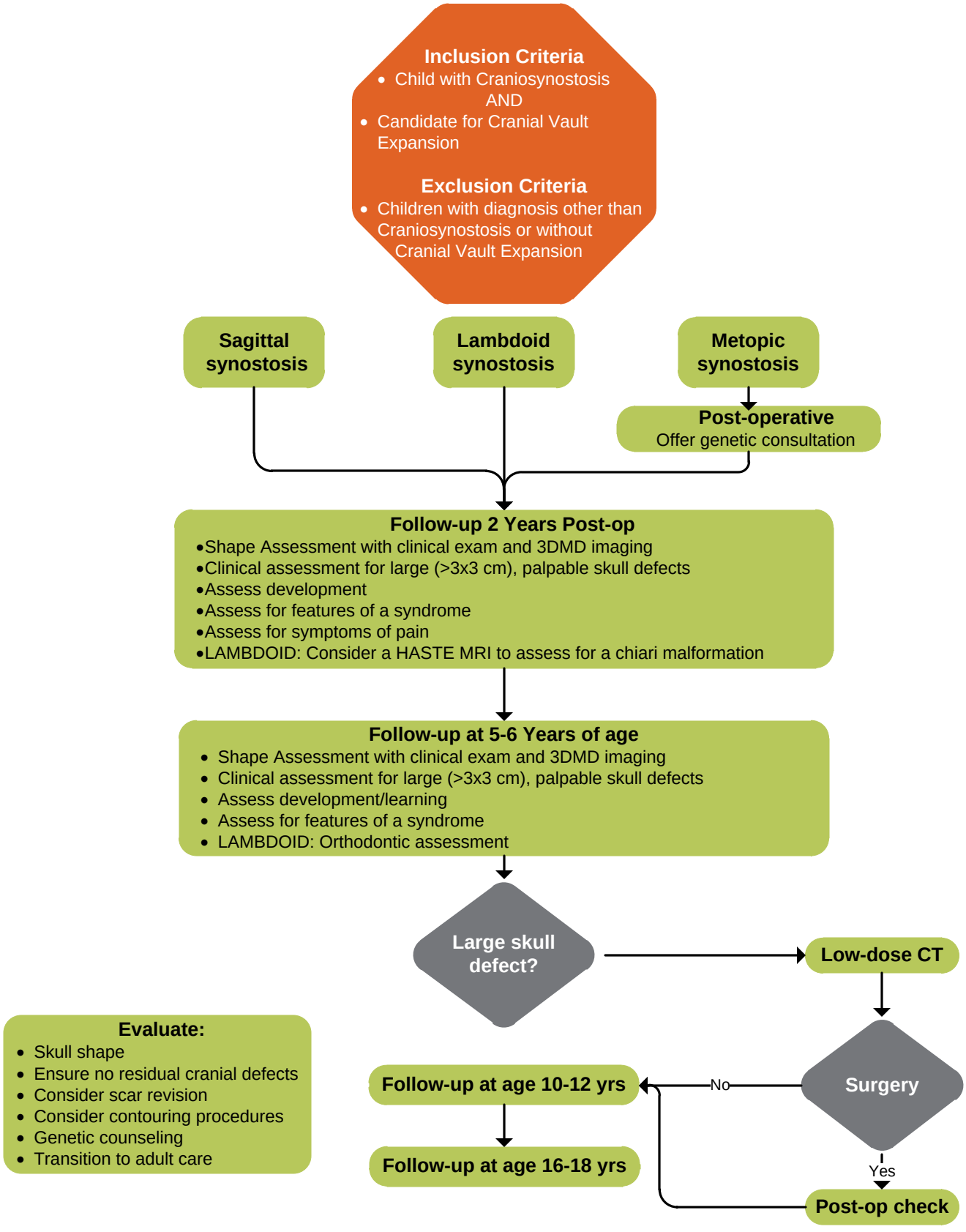
Craniosynostosis v4.0: Long Term Follow-up Sagittal, lambdoid, Metopic

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

Single Suture Craniosynostosis: Sagittal, Lambdoid, Metopic



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Craniosynostosis v4.0: Long term follow-up Coronal

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

Coronal Craniosynsotosis

Uni/Bi-Coronal (includes Muenke, Saethre Chotzen Syndrome, and apparent isolated coronal synostosis

Inclusion Criteria
- Child with Craniosynostosis AND
- Candidate for Cranial Vault Expansion

Exclusion Criteria
- Children with diagnosis other than Craniosynostosis or without Cranial Vault Expansion

CVR typically involves FOA age 12 months

1st Visit
- Screen for sleep-disordered breathing
- CT scan with c-spine - head and face through c- spine
- Offer genetic counseling visit and testing- preauthorization order, DNA banking
- Standardized statements in notes

Hearing evaluation in the first year

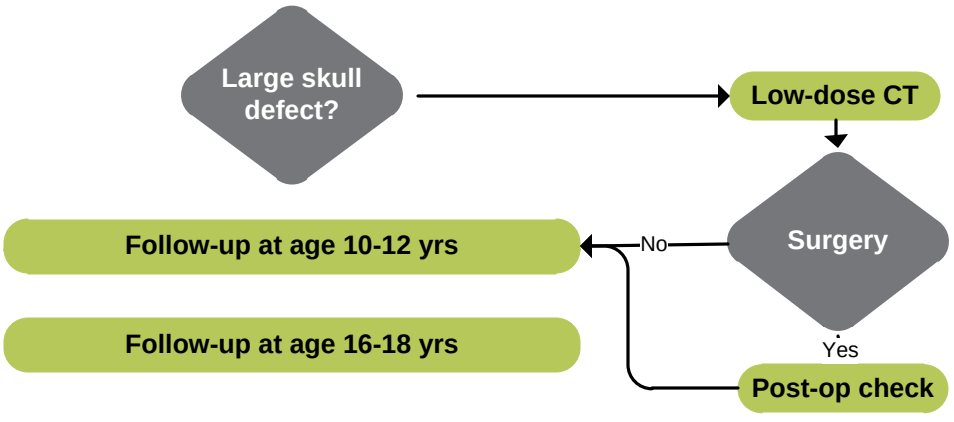
Surgery
CVR as indicated may need more than one procedure during the first 2 years

Post-operative
Ophthalmology assessment to rule out strabismus

Follow-up 2 Years Post-op
- Shape assessment with clinical exam and 3dMD imaging
- Clinical assessment for large (>3x3 cm), palpable skull defects
- Assess development
- Assess for features of a syndrome
- Assess for symptoms of pain
- Repeat audiogram
- Assess for cervical spine anomalies, if not already done with pre-op CT
- Ophthalmology follow up yearly until 6

Follow-up at 5-6 Years of age
- Shape Assessment with clinical exam and 3dMD imaging
- Clinical assessment for large (>3x3 cm), palpable skull defects
- Assess development/learning
- Assess for features of a syndrome
- Screen for sleep-disordered breathing
- Orthodontic evaluation

Evaluate:
- Skull shape
- Ensure no residual cranial defects
- Consider scar revision
- Consider contouring procedures
- Genetic counseling
- Transition to adult care



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Last Updated: March 2019
Next Expected Review: March 2024

Craniosynostosis v4.0: Crouzon / Pfeiffer Syndrome

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

Crouzon / Pfeiffer Syndrome

As soon as the syndrome has been identified, these things should be done:
- Airway evaluation/OSA Screen
- Diagnostic hearing test
- Offer genetic counseling and testing
- NDV referral at age 2 years
- Early intervention ASAP

Inclusion Criteria
- Child with Craniosynostosis AND
- Candidate for Cranial Vault Expansion

Exclusion Criteria
- Children with diagnosis other than Craniosynostosis or without Cranial Vault Expansion

Surgery
- CVR as indicated may need more than one procedure during the first 2 years

*Need to identify congenital and progressive cranial and vertebral fusions, obtain 4V cervical spine films – age 4-5 years

Annual visits between ages 2-5 years
- Ophthalmology assessment*
- Developmental assessment
- Speech assessment (if a cleft palate had not already been identified)
- Assess for cervical spine anomalies. if screening c-spine CT is normal perform follow-up c-spine evaluation at age 4-5 unless clinically indicated sooner
- Airway assessment
- Hearing assessment
- Assess for a chiari malformation with a full MRI* at age 4 years, if not already done. Consider combining with another procedure- before mid-face surgery
- Clinical assessment for large (>3x3 cm), palpable skull defects
- Assess for cervical spine anomalies if not already done with pre-op CT

**MRI brain and screening sagittal MRI of spine. If radiologist sees a syrinx, then will request a full spine.

Annual visits between ages 6-15 years
- Orthodontics assessment
- **Consider** LF III between age 6-10 years, recommend pre-op CT with angiogram
- Ophthalmology follow up
- Developmental follow up
- Airway re-assessment
- Audiology follow up

Annual visits between ages 16-21 years
- Consider final orthognathic surgery
- Follow up with assessments described above
- Genetic counseling
- Transition to adult care

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Craniosynostosis v4.0: APERT SYNDROME

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

APERT SYNDROME

Upon diagnosis:
- Initial team evaluation: PS, NSR, OTO, Peds, RN, SW, Audio
- Within the first 6 months: hand, ophtho- timing of f/u for synonychia, syndactyly?
- Determine timing of CT Scan, CVR
- With first intubation, combine case with OTO for intra-operative airway evaluation
- Genetic counseling visit in the first year
- Early intervention referral at diagnosis
- NDV referral in first 2 years
- Peds/RN determines frequency of visits in the first 2 years

Inclusion Criteria
- Child with Craniosynostosis AND
- Candidate for Cranial Vault Expansion

Exclusion Criteria
- Children with diagnosis other than Craniosynostosis or without Cranial Vault Expansion

Common CVR procedures include:
- PVDO age 6-12 mos (rarely neonatal)
- FOA age 18-24 mos

Hand Surgery
- Plan hand surgery around CVR, not combined
- Surgical interventions typically occur between 6 mos-2 years

Surgery
- CVR as indicated may need more than one procedure during the first 2 years

Yearly between ages 2-5
- Ophthalmology assessment*
- Developmental assessment*
- Speech assessment (earlier if cleft palate was already identified)
- Assess for signs of hydrocephalus, request NSR and imaging as needed
- Assess for cervical spine anomalies. If screening c-spine CT is normal, perform follow-up c-spine evaluation at age 4-5 unless clinically indicated sooner
- Vertebral spine assessment
- Airway assessment
- Audiology and ENT assessment for eustachian tube dysfunction
- Orthopedic hand follow up
- Clinical assessment for large (>3x3 cm), palpable skull defects

Yearly between ages 6- 10
- Orthodontics assessment*
- **Consider** LF II/III between age 6-10 years, recommend pre-op CT with angiogram

Yearly between ages 11-21 years
- Consider final orthognathic surgery
- Start transition to adult care. Consider UW transition program (if applicable), identify subspecialty needs

For questions concerning this pathway, contact: craniosynostosis@seattlechildrens.org

Last Updated: March 2019
Next Expected Review: March 2024

Craniosynostosis Approval & Citation

Approved by the CSW Craniosynostosis for March 28, 2019 go-live

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Executive Approval:

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Surgeon-in-Chief	Bob Sawin, MD

Retrieval Website: http://www.seattlechildrens.org/pdf/craniosynostosis-pathway.pdf

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Return to Home

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Evidence Ratings

This pathway was developed through local consensus based on published evidence and expert opinion as part of Clinical Standard Work at Seattle Children's. Pathway teams include representatives from Medical, Subspecialty, and/or Surgical Services, Nursing, Pharmacy, Clinical Effectiveness, and other services as appropriate.

When possible, we used the GRADE method of rating evidence quality. Evidence is first assessed as to whether it is from randomized trial or cohort studies. The rating is then adjusted in the following manner (from: Guyatt G et al. J Clin Epidemiol. 2011;4:383-94.):

Quality ratings are *downgraded* if studies:

- Have serious limitations
- Have inconsistent results
- If evidence does not directly address clinical questions
- If estimates are imprecise OR
- If it is felt that there is substantial publication bias

Quality ratings are *upgraded* if it is felt that:

- The effect size is large
- If studies are designed in a way that confounding would likely underreport the magnitude of the effect OR
- If a dose-response gradient is evident

Quality of Evidence:

✪✪✪✪ High: The authors have a lot of confidence that the true effect is similar to the estimated effect
✪✪✪○ Moderate: The authors believe that the true effect is probably close to the estimated effect
✪✪○○ Low: The true effect might be markedly different from the estimated effect
✪○○○ Very low: The true effect is probably markedly different from the estimated effect
Guideline: Recommendation is from a published guideline that used methodology deemed acceptable by the team
Expert Opinion: Based on available evidence that does not meet GRADE criteria (for example, case-control studies).

Return to Home To Bibliography

Summary of Version Changes

- **Version 1.0 (08/08/2012): Go live**
- **Version 2.0 (05/28/2014): Changes to Inclusion/Exclusion criteria to reflect single and multi suture Craniosynostosis**
- **Version 3.0 (12/16/2016): Added Cervical CT for patients with syndromic synostosis**
- **Version 4.0 (3/28/2019): Minor modifications to phase I, with the addition of phase II post op and outpatient care.**

Return to Home

Medical Disclaimer

Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in treatment and drug therapy are required.

The authors have checked with sources believed to be reliable in their efforts to provide information that is complete and generally in accord with the standards accepted at the time of publication.

However, in view of the possibility of human error or changes in medical sciences, neither the authors nor Seattle Children's Healthcare System nor any other party who has been involved in the preparation or publication of this work warrants that the information contained herein is in every respect accurate or complete, and they are not responsible for any errors or omissions or for the results obtained from the use of such information.

Readers should confirm the information contained herein with other sources and are encouraged to consult with their health care provider before making any health care decision.

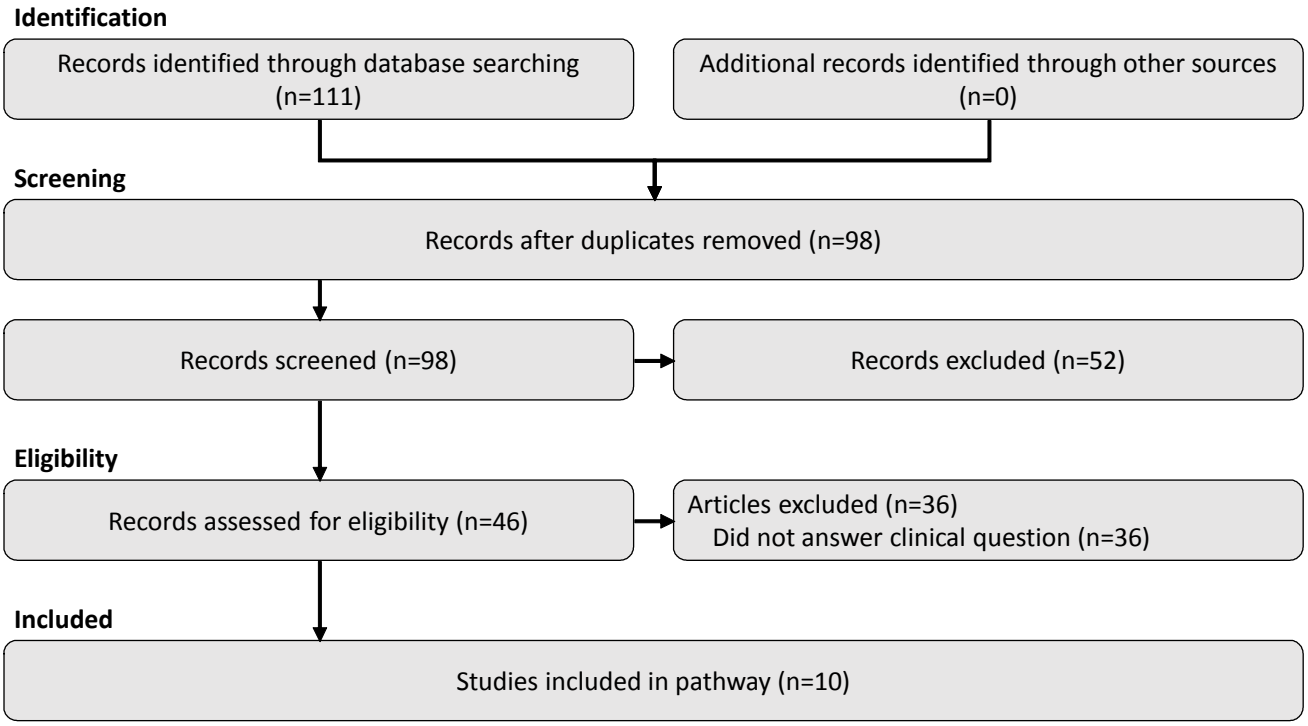
Return to Home

Bibliography

CSW Craniosynostosis Methods

Literature searches were conducted in June of 2018. The search targeted synthesized literature on craniosynostoses, including related sysnostoses, cranial vault, cranial sutures or plagiocephaly. The search was conducted for 2012 to current and limited to English. The search was executed in Ovid Medline, Embase, Cochrane Database of Systematic Review, National Guidelines Clearinghouse and the Turning Research into Practice database (TRIP) databases.

Jackie Morton, MLS
March 5, 2019



Return to Home
To Bibliography, Pg 2

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