Children's Hospital of Pittsburgh of UPMC

CRANIOSYNOSTOSIS PROTOCOL

THE PRIMARY ROLE FOR OPHTHALMIC SURVEILLANCE IS TO PREVENT VISUAL LOSS
Strabismus in these patients is anomalous and each child must be assessed by the orthoptist. All decisions regarding surgery for decision MUST be consultant led.

Syndromic

Apert, Crouzon, Pfeiffer : Allelic Disorders – Mutations in FGFR2
Saethre-Chotzen : TWIST gene mutatio
CranioFrontoNasalDysplasia

Evaluation by Orthoptist, Electrophysiology, **Genetic counselors** and Dr's team
In addition to routine eye exam **MUST HAVE**

- Dilated fundoscopy at every visit if under 8 years of age
  Optic disc images if possible
- Monitor ocular surface treat as required
  - Lubricants
  - Temporary lateral tarsorraphy as a short term measure. Observe IOP carefully

Risk of scarring and reduced visual acuity if untreated

Non-Syndromic

Every new referral should be seen by Orthoptist and Dr's team and Genetic counselors

- Sagittal , Bicoronal
  - Electrophysiology
    - Review
      3-6 monthly until 4 yrs
      6-9 monthly until 8yrs
      Shared care after 4 yrs
- Unicoronal
  - 3-6 monthly until 4yrs
    6-9 monthly until 6 yrs and then discharge/shared care
    - AHP in unicoronal
      Base of skull asymmetry which may be the cause
      May be SO palsy

Treat amblyogenic factors as in any other patient (see amblyopia protocol)
CAVEATS : Patches may not WORK
-Consider atropine penalisation
-Ensure cyclopegic refractions at least 6 monthly if under 8yrs
-Dispense glasses with consideration for facial features
-Occlusion therapy as indicated – see above

Concerns over raised ICP

- Swollen discs – raised ICP til proven otherwise
  ASK ABOUT
  Sleep apnoea – snorinG
  Adenoid-tonsillectomy
  On CPAP
  Behavioural changes
- Abnormal VEPS with swollen discs – raised ICP until proven otherwise
  Abnormal VEPs but no swelling and no other concerns review 6-8/52 with repeat electrophysiology
- Ensure craniofacial team aware of our concerns so that other contributing factors may also be identified/managed
  AirwaY,CT Scan, etc

Review
2-3 months up to 4yrs age

4-6 months once over 4 years if all stable and no concerns

Once 8 yrs very rare to see raised ICP.
9monthly review

This clinical guideline is a collaborative care plan and is not intended to construed or to serve as a standard of medical care. Rather, it is intended as a guideline to promote coordination and communication with respect to patient care and may be modified to meet individual care needs. For additional information please contact the Department of Clinical Effectiveness & Quality at 412/692-7645. ©Children's Hospital of Pittsburgh of UPMC, 2014