Surgery: Generalized Epilepsy/Corpus Callosotomy

Francesco T Mangano, DO, FACS FAAP
Mary Jane and Bob Tritsch Professor of Neurosurgery & Pediatrics
Cincinnati Children's Hospital Medical Center
University of Cincinnati College of Medicine, Cincinnati, Ohio

I. Generalized Semiology:

1. Atonic Seizures:
    - Synchronous voluntary muscle tone increase followed by loss of resting tone. Considered inhibiting-suppressing resting muscle tone
    - Patients are at significant risk for polytrauma.
2. Four Clinically distinct subtypes:
    - Increase in axial and appendicular tone prior to atonic phase.
    - Forward flexion of neck, shoulders and flexion of thighs.
    - Trunk dorsi-flexion and arm straightening.
    - Pure atonic-rare but best prognosis.
3. Most occur in the morning after arousal from sleep and in brief episodic clusters.
4. Lennox-Gastaut Syndrome: Triad
    - Mixed atonic seizures
    - Cognitive delays
    - Abnormal EEG tracing with interictal slow spike and wave discharges.

II. Associated EEG:

1. Ictal EEG: Generalized spike and wave, polyspike discharges of rhythmic slow waves and fast recurring rhythms.
2. Interictal EEG: Poorly organized with bilateral synchronized spike and wave
    - Complexes 1-2.5/sec in bursts lasting seconds to minutes.
    - Must differentiate from 3/sec spike and wave of typical petit mal absence seizures.

III. Medical Management

- AED Management is disappointing usually refractory to monotherapy.
- Typical AEDs include phenobarbital, valproic acid, and ethosuximide.
- Generally valproic acid has historically worked best.
- Ketogenic diet can be considered for children younger than 6 years of age.

- Environmental modifications help with potential injuries:
    - Helmet
    - Carpeted floors
    - Removal of sharp objects from the environment

IV. Surgical Management

- Palliative procedures include sectioning of corpus callosum and vagal nerve stimulator therapy.
- Presumptive mechanisms: disruption of transhemispheric connections and synchronization of epileptic discharges.
- Corpus Callosotomy first developed by van Wagenen in 1940.
- Indicated for those with partial seizures with rapid generalization without identifiable focus, and refractory synchronized seizures.

V. Surgical Procedure:

- May be performed as partial (Ant 2/3) or complete.
- Anterior 2/3 corpus callosotomy: Rostrum to isthmus
- Complete corpus callosotomy: Rostrum to splenium.
- May be performed through an open craniotomy approach or endoscopic approach.
- Benefits of CCC: Better seizure control, higher likelihood of complications
- Benefits of Endoscopic Approach: Minimally invasive, less brain retraction, less post-operative pain, steep learning curve.

VI. Open Craniotomy for Corpus Callosotomy:

- Patient positioning- Supine or lateral in 3-Point fixation.
- Incision may be bicoronal or trap-door type.
- Rectangle craniotomy off midline, generally right side, crossing coronal suture.
- Burr hole for craniotomy are placed on or close to sagittal suture and 4cm lateral to midline.
- Care needs to be taken to avoid bleeding over superior sagittal sinus (SSS).
- Risk of venous air embolism.
- Dura is open with base on SSS. Interhemispheric tissue is identified.
- A retraction system is used to retract the medial frontal lobe laterally.
- Interhemispheric dissection splits arachnoid connections caudally to the pearly white CC.
- Using high magnification the distal anterior cerebral arteries (pericollosal) are identified and retracted to provide a safe corridor in the mid- corpus callosum.
- The callosotomy starts along the mid body of CC and extends anteriorly around the genu to the rostrum.

VII. Complete Corpus Callosotomy:

- The midline of the CC is identified by gentle aspiration while looking for the ependyma of the lateral ventricles as it extends over the leaves of the midline septum. This is a key finding that then allows one to stay in the midline as the procedure is completed from the rostrum to the isthmus (Ant 2/3) or to complete the cc to the posterior splenial margin of the CC.
- Posterior resection is completed when the pia overlying the vein of Galen is clearly identified.
- Attempt is made not to enter the ventricles as this creates the potential of a post-op CSF leak, impaired visualization, and can cause intracranial shift.
- Once the callosotomy is complete, the field is thoroughly irrigated of any serosanguinous fluid and the intracranial retractors are removed. The dura is closed and the craniotomy is replaced with titanium screws and plates to allow healing. The scalp is closed with sutures.

VIII. Alternate Approaches

- Endoscopic
  - Adjuncts: neuronavigation
  - Incision linear, lateral to midline
  - Positioning supine, head elevated slightly
  - D shaped craniotomy with base along SSS
  - Dural opening with SSS Base
  - May get relaxation of brain after dural opening with external ventricular drain or mannitol dosing
  - The endoscope is used to dissect the interhemispheric fissure and separates the cingulate gyri.
  - A combination of ultrasonic aspirator and endoscope are used to disconnect the corpus callosum
  - Same principles of hemostasis, careful retraction around ventricles, avoidance of CSF leak, watertight dural closure, replacement of craniotomy flap and tissue closure apply.
- Laser Ablation
  - Emerging technique
  - Requires multiple trajectories to ablate the complete corpus collosum
  - Performed in intra-op MRI scanner
  - Utilizes heat sink from blood vessels and CSF to avoid inadvertent injury to other anatomic structures
- Radiosurgery for Callosotomy
  - Described in the literature
  - Not the standard but appropriate for selected cases

IX. Outcomes

- Single stage CCC appears to have better surgical outcomes with lesser risk of neurological and operative complications
- Anterior 2/3 disconnection and 2-stage completion of CC and single stage CCC all result in varying degrees of resolution of absence, astatic, myoclonic, GTC and complex partial seizures.
- Complications include:
    - Disconnection Syndrome- usually subsides within 2-3 weeks; in adults it may take as long as 2 years to improve
    - Hemiparesis-akinesia
    - Speech abnormalities- mutism
    - Surgical Complications
        - Infection
        - Hemorrhage EDH/SDH
        - CSF leak
    - Younger patients may have better outcomes
- Drop Attacks: Seizure freedom 84% (6 year follow up)
- GTC: 27 %
- Absence 31%
- Complex Partial 14%
- Simple Partial 40%

X. Callosal Disconnection Syndrome

- Reduced by Ant. 2/3 callosotomy
- Variable expression of symptoms for interrupted connections between hemispheres
    - Symptoms:
        - Motor control-apraxia left sided
        - Spatial orientation
        - Vision
        - Hearing
        - Language-aphasia
    - <u>Acute</u>
        - SMA-like syndrome more prominent and prolonged with CCC.
        - Paresis of non-dominant leg, incontinence, delay of speech. When severe patients may be unresponsive to environment although awake. Lasts days to months.
        - Nearly always resolves.

- Chronic
    - Moderate memory deficits with difficulty with uncommon paired words, topographical memory.
    - Precentral gyrus disconnection: Alien Hand Syndrome.
    - Temporal lobe disconnection: Inability to differentiate auditory stimuli between dominant and non-dominant input.
    - Postcentral gyrus disconnection: tactile dysnomia.
    - Parietal lobe disconnection: language difficulty necessitating visual frame of reference. Alexia without agraphia.
    - Occipital lobe disconnection: - sensory disconnection syndrome.

References

Corpus callosotomy-Open and endoscopic surgical techniques. Smyth MD, Vellimana AK, Asano E, Sood S. *Epilepsia*. 2017 Apr;58 Suppl 1:73-79. Review.

Corpus callosotomy outcomes in pediatric patients: A systematic review. Graham D Tisdall MM, Gill D. *Epilepsia* 2016 Jul;57(7):1053-68.

Outcomes after anterior or complete corpus callosotomy in children. Kasasbeh AS, Smyth MD, Steger-May K, Jalilian L, Bertrand M, Limbrick DD. *Neurosurgery*. 2014 Jan;74(1):17-28.

Long-term follow-up of seizure outcomes after corpus callosotomy. Sunaga S, Shimizu H, Sugano H. *Seizure*. 2009 Mar;18(2):124-8.

David W Roberts and Adrian Seigel (2001) Corpus Callosotomy. Eds. Luders and Comair in *Epilepsy Surgery* (82) pp747-756. Lippincott Williams & Wilkins, Philadelphia PA.

James Leiphart and Itzhak Fried (2011) Surgery for extratemporal lobe epilepsy. Ed. Winn H.R. *In Youman's Neurological Surgery* Vol. 1:(61) Pp. 759. Elsevier Saunders, Philadelphia PA.

Atonic Seizures. Duchovny M. *Pediatrics in Review 1987;*9;2;43.

Corpus callosotomy in children and the disconnection syndromes: a review. Jea A, Vachhrajani S, Widjaja E, Nilsson D, Raybaud C, Shroff M, Rutka JT. *Childs Nerv Syst*. 2008 Jun;24(6):685-92.