Epilepsy and epilepsy surgery in tuberous sclerosis

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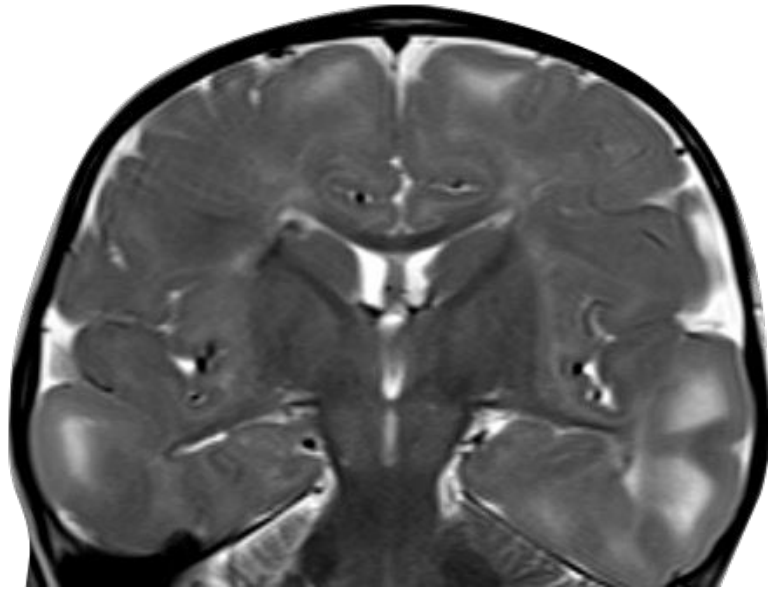
Epilepsy in tuberous sclerosis complex (TSC)

- seizures occur in 60-90% of children with TSC
  - the very first symptom is a seizure in 70% of children
  - most seizures start in the first year of life as a baby
- seizure types
  - >90% have focal seizures
  - 50% have epileptic spasms, associated with developmental regression/delay
- seizures not controlled with medication in >50% of children

Ihnen SKZ, *Epilepsia* 2025

Learning difficulties & epilepsy

- learning difficulties, intellectual disability and autism are common in tuberous sclerosis
  - ~ 50%
- the severity of learning and behavioural difficulties is strongly correlated with epilepsy severity
  - early onset seizures
  - uncontrolled seizures
  - infantile spasms
  - high seizure burden
  - drug-resistance



Chu-Shore CJ, *Epilepsia* 2010; Yates JRW, *Arch Dis Child* 2011; Curatlo P, *Lancet Neurol* 2008; Staley BA, *Pediatrics* 2011; Humphrey A, *Epilepsia* 2014; Mitchell R, *DMCN* 2021; Ihnen SKZ, *Epilepsia* 2025,

Why perform epilepsy surgery in children with TSC?

Reduce or stop seizures

May improve learning / cognitive trajectory

Come off or reduce medications

Reduce clinic visits, hospital admissions

Decrease risk of injury from seizures

May prevent more “severe” epilepsy like Lennox Gastaut syndrome

Decrease risk of sudden unexplained death in epilepsy

When should epilepsy surgery be considered in TSC?

- At the RCH we evaluate any child with TSC who has:
  - drug-resistant focal epilepsy = tried 2 or more antiseizure medications and has ongoing seizures
  - infantile or epileptic spasms which have not resolved with steroid treatment
- Children should be urgently evaluated if they are young and/or have developmental delay or regression
- No requirement to have tried other therapies first (e.g. ketogenic diet)

Benefits of early surgery in drug-resistant epilepsy

- TimeToStop study:
  - study looking at when is best to stop antiseizure medications
  - earlier and complete medication withdrawal was associated with improved postop IQ scores
- seizure freedom and stopping antiseizure medications shown to be associated with improved developmental outcomes

Boshusein et al. 2015 *Annals of Neurology*, Eriksson et al. 2024 *Brain* Stefanos-Yakoub et al. 2023 *Epilepsia*

Surgery goals in TSC are different to other patients who have epilepsy surgery

- the goal is not always complete seizure freedom
  - other tubers & genetic variant may later cause seizures
- the goals are:
  - reduce seizure frequency
  - reduce the burden and side effects of being on multiple medications
  - improve development & the opportunity to “be a kid”
  - avoid progression of epilepsy to a developmental and epileptic encephalopathy or Lennox Gastaut syndrome
    - where the epilepsy itself impacts a child’s learning and behaviour

What is the “pre-surgical” evaluation

- Goals
  - find the area to operate on, e.g. epileptogenic tuber
  - determine how safe it is to operate in that area
- Process
  - Clinical history
    - detailed questions about what happens during the seizure / sequence of events, how many seizure types, when seizures occur, how often they occur
    - medications tried
    - developmental impact
  - Examination
  - Review of videos – very helpful to see videos across time

What is the “pre-surgical” evaluation

- Process (continued)
  - Video-EEG monitoring
    - +/- electrical source imaging
  - MRI brain
  - +/- FDG-PET
  - Developmental assessment or neuropsychology
- Discussion in multidisciplinary meeting:
  - epileptologists, neurosurgeons, radiologists, EEG scientists, neuroimaging scientists, psychologists
- Feedback from MDT and recommendation from surgery or not to family

Why is (sometimes) difficult about surgical planning for the neurologist with TSC?

- often difficult to know which tuber is causing the seizures
  - e.g. compared to a child who has a different condition with a single brain lesion
- often doing surgery at a young age
  - child unable to tell us about their seizure
  - MRI immature
  - unable to go some of the “usual” presurgical planning tests, e.g. language fMRI scans
- sometimes EEG findings are difficult to interpret, e.g. infantile spasms, immature EEG

What happens on the day (at RCH)?

- general anaesthetic
  - intubation
  - lines – central line, arterial line (measure BP), venous lines
  - MRI scan
- preparation by surgeon
  - shaves area of hair
  - opens skin (often long incision in hair line)
  - removal of bone
- EEG recordings
  - to see what the brain looks like before surgery
  - special anaesthetic
- surgery
  - removal of epileptic tuber or tubers
- EEG recordings +/- further resection
  - to determine if spikes have gone

What is the recovery process?

- in hospital
  - most children do not require ICU admission following surgery, nursed on the ward
  - maximum swelling at 48 hours, day 2 postop often the hardest, ”black eye”
  - discharge usually by day 4-5
- at home
  - may need more rest
  - no sports/wrestling/rough play 8 weeks, no swimming 4 weeks
  - gradual return to school from week 2-3 post-op

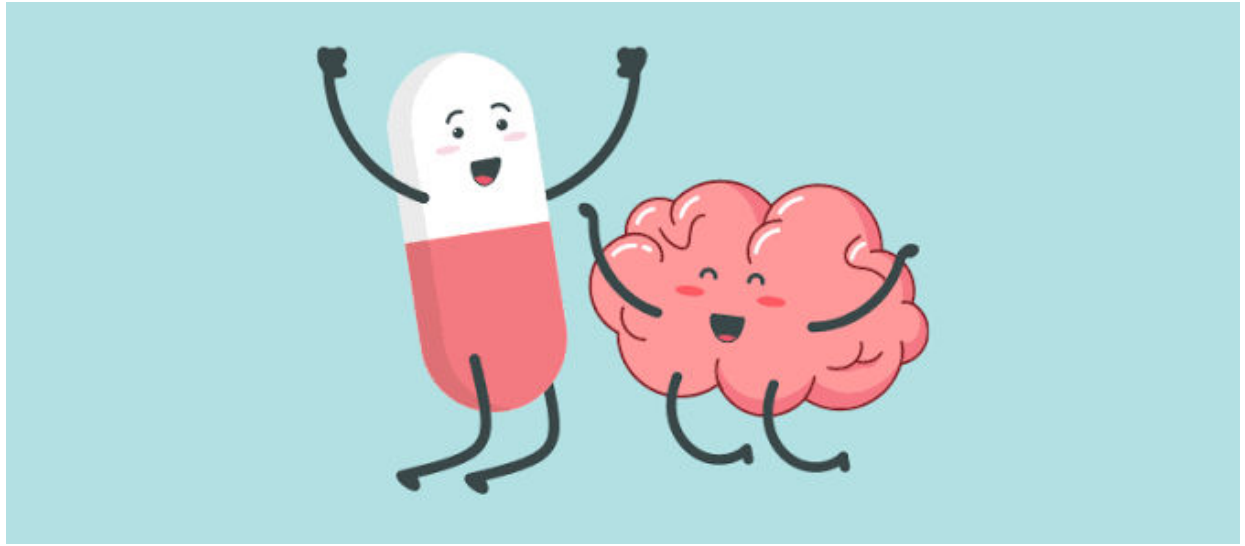
RCH experience

- 99 children, 173 operations since we started operating on kids with TSC (1997)
- all tuber or tuber centre resections
- almost 90% younger than 3 years
- pre-op
  - all had drug resistant epilepsy
  - 66% with spasms +/- focal seizures
  - 33% focal seizures only
- at follow-up:
  - 60% completely seizure free
  - 35% ongoing focal seizures
  - 5-10% ongoing spasms
  - 20% normal intellect

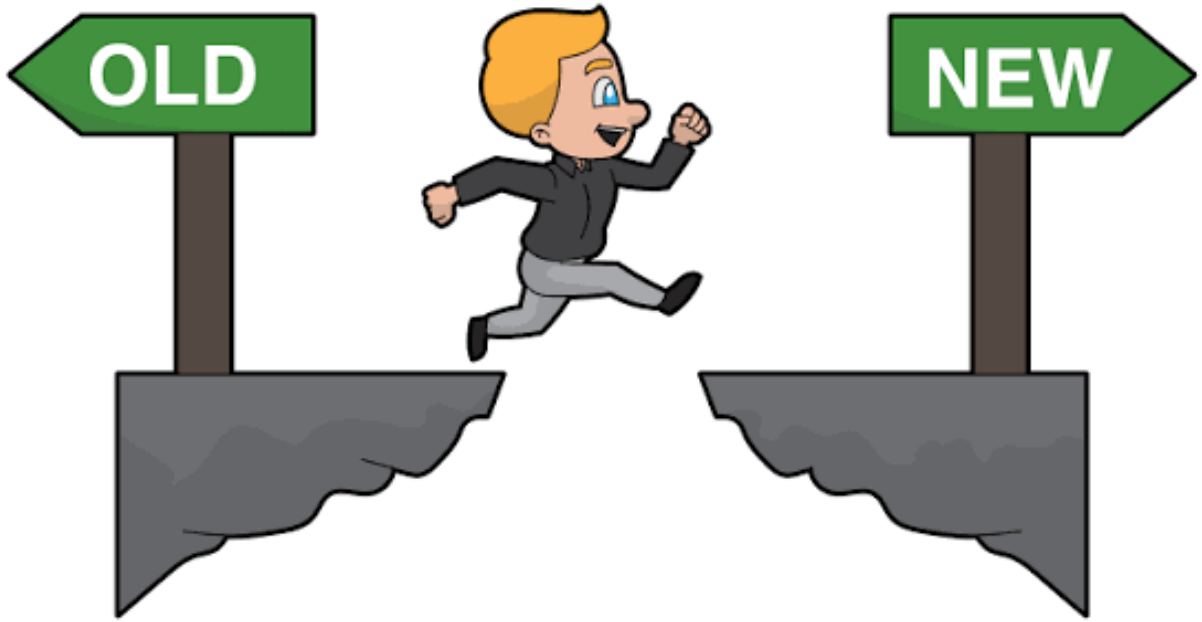
Questions from parents

Is making the decision to do surgery in my child's best interests?

Will the surgery be painful? How painful?



What if my child is completely different after surgery?



What if my child has a complication?

What if the surgery doesn’t stop seizures? Will more surgeries be required?

Advice from other families

Get a good team, ask questions

- relationship with medical & surgical teams is important
- use medical resources available
  - epilepsy nurse, nurse specialist
  - social worker
  - general practitioner
  - paediatrician
  - psychologist
  - Tuberous Sclerosis Australia
  - Epilepsy Foundation, Epilepsy Australia
  - families who have been through epilepsy surgery
- home team
  - family, neighbours, friends
  - school

Plan what you will do on the day

- best to leave the hospital if possible, during surgery
- plan who will stay with child, who is looking after other family members, shifts
- transport to and from hospital, accommodation if necessary
- clothes, toys, extra food, low stimulus activities

Look after yourself, marathon not a sprint

- Ensure you are doing the basics:
  - eating, hydration, sleep, exercise, getting outside
- Lean on supports
- Hospitals are difficult – disruption, lack of information, changes in plans
  - ask questions – nursing staff first point of call
  - escalate if you are worried – most hospitals have patient-initiated codes now

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