



The Tuberous Sclerosis Association

This document can be found at www.tuberous-sclerosis.org

It reports on a presentation at a meeting of the TS Alliance in San Diego in July 2001.

Beyond Anti-Convulsants – Epilepsy Surgery, VNS and Ketogenic Diet

Surgery for Seizures in TS – Dr Peter Black, Neurosurgeon at Harvard Medical School.

The dysplastic brain in TS is responsible for the epilepsy and this part of the brain can be identified using imaging and mapping techniques. The challenge with epilepsy surgery in TS is to select the right tuber, avoiding other important bits of the brain and then weighing the risk/benefit ratio first.

Selecting the correct tuber involves imaging the brain, performing EEGs, undertaking long-term monitoring with scalp electrodes, and invasive monitoring to correlate the epileptic activity with the relevant tuber.

Avoiding important bits of the brain involves Transcutaneous Magnetic Stimulation (TMS) and a functional MRI. The risk-benefit ratio is a team decision which takes into account the family and the patient's wishes, the chance of seizure improvement, the severity of the present situation (including drugs) and the risk of harm. There is a risk of polypharmacy on the developing brain, a risk which we may underestimate.

TMS involves inserting grids under the scalp and placing them over the top of the brain. It's possible to stimulate individual ones to test reactions or response. The surgeon can overlay this grid with an MRI 3 dimensional map of the structure of the brain and see which bit of the brain is affected when individual parts of the grid are stimulated with a very gentle current under local anaesthetic while the patient is still awake. It is possible to dissect a tuber via the sulcus while the patient is still talking. He predicted the possibility that the future of brain surgery lay in the brain scanner with imaging as you go!!

Perhaps half of children with TS have focal seizures. Of a group of 14 TS children who've had epilepsy surgery to remove a tuber, 13 had a marked improvement in their seizure frequency after surgery and 1 had an increase in seizures.

Dr Black looked forward to a new era of epilepsy management through surgery, with the biggest challenge being to identify the offending tuber and remove it safely.

Ketogenic Diet

Hippocrates noticed that a man had his seizures completely controlled by having no food and drink, and Jesus had cast out demons from epileptic children through fasting and prayer. Ketosis means a fasting state, when the body uses glucose, glycogen and then breaks down fats. The result is an excess of ketones. Ketones are used by the brain as an alternative source of energy. There was a resurgence of the ketogenic diet in 1994.

The typical American diet involves 50% carbohydrates, 45% fat and 15% protein. The Ketogenic diet is 6% carbohydrate, 86% fat and 8% protein. It involves a restriction in calories and fluid, too, and supplements (minerals and vitamins) are necessary. How it works is unknown, but sometimes it can result in more than a 50% reduction in seizures. It is used in drug-resistant epilepsy or when there are undesirable medication side effects. But the diet has side effects, too – hypoglycaemia, hypocalcaemia, acidosis, decreased appetite, hyperlipidemia, hyperurecemia, hypercalcuria, cardiac failure.

3 meals and 2 snacks are taken per day. The food is weighed and measured and various supplements are given. A seizure reduction should be seen in the first month,

Vagal Nerve Stimulation

By 2001, over 10,000 patients have been treated with vagal nerve stimulation world-wide. It has been used as an add-on therapy in those over the age of 12 with partial onset epilepsy. The exact mode of action is unknown. There are possible 2 responses to VNS. One is immediate, one is over a period of time. PET scanning studies have, over time, shown changes in blood flow in the brains of patients who've had VNS. Improvement continues over time. Nearly 50% have had more than 50% reduction in their seizures over time following VNS. Very few are seizure free. Side effects include hoarseness, cough, paresthenia, dyspnea, scarring and voice change. Side effects decline with time. Quality of life is better – people are more alert, less post-ictal, less moody, with an improved memory.

20% TS patients aged 12 and over had a greater than 90% reduction in seizure frequency over one year. It has been suggested that the earlier the treatment is given, the better the outcome. It is likely that there will be a dramatic improvement in drop attacks and tonic seizures. It has also been discussed as a use for depression.