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BRAINCHILD

The Official Publication of HKCNDP

Special Issue on Paediatric Neurophysiology Update by the British Columbia Children's Hospital Team

香港兒童腦科及體智發展學會

The Hong Kong Society of Child Neurology and Developmental Paediatrics

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Our appreciation of thanks to Cheung Man Chun and Ng Sum Lui (students of Hong Kong Christian Service Kwun Tong Nursery School) for the cover drawing named "Magic Forest".

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The Hong Kong Society of Child Neurology and Developmental Paediatrics

EDITOR'S NOTES for the July 2016 Issue

Paediatric Neurophysiology Update by the British Columbia Children's Hospital Team

Dr. Chok Wan CHAN

The current issue of Brainchild is devoted to "Paediatric Neurophysiology Update" capably edited by Professor Peter K. H. Wong B. Eng MD FRCP(C), Professor for Division of Paediatric Neurology, Department of Paediatrics, University of British Columbia, Vancouver, Canada and Director of Department of Diagnostic Neurophysiology, Children's Hospital, Vancouver, B.C., Canada. Through his expert team members, we are able to appreciate the modern team approach to neurophysiology and the multidisciplinary, interdisciplinary and transdisciplinary collaboration in the neurosciences. Thanks to members of the team and we take great pride to present this comprehensive update to all professionals in neurosciences consisting of doctors, nurses, allied health professionals, social workers, psychologists, special teachers and educated patients for their comments and positive feedbacks. We are grateful to Professor Wong for his great leadership and to all the authors for their effort. We appreciate your contributions to the child health community!

Year 2016 is the 22nd anniversary for the Hong Kong Society of Child Neurology and Developmental Paediatrics (HNCNDP). We are pleased to witness our achievements and on looking back we can summarize our activities in the past two decades into several major channels:

1. Regular Professional activities: we have bimonthly meetings on each of the following:
   a. Child Neurology Conference
   b. Neuro-developmental Conference
   c. Regular Bimonthly Scientific Meetings
2. Annual Scientific Meeting: each year we invite a world expert on one aspect of either Developmental Behavioural Paediatrics (DBP) or Child Neurology (CN) from Untied States of America, Canada, United Kingdom, Europe, Australia and other countries to deliver a series of state-of-art aspects of the subject and through participation of local professionals we successfully hosted a significant number of symposia, seminar, workshops and fora on the various aspects of the subject. The climax is the Keynote Lecture and Dinner each year on the themed topic which remains one of the most popular scientific activities attracting hundreds of professionals each year. So far we have hosted more than 20 such annual scientific meetings. Readers can refer to the Society website for more details.
3. Publications - the Society launched regularly the following publications consistently, professionally and academically:

a. The Brainchild – this publication is devoted to special issues on the various aspects of the DBP and CN and is distributed to healthcare professionals and professionals dedicated to neurosciences free of charge. They are available in most of the University Libraries and the public libraries for health educations.
b. The Society Newsletter – this is meant to circulate to professionals on recent advances, meeting announcements and social activities.

4. Professional Accreditations : the Society professionally assists the Hong Kong College of Paediatricians in accrediting subspecialists in CN and DBP via the establishment of two Subspecialty Boards on Child Neurology and on Developmental Behavioural Paediatrics for:
   a. Accreditation of First Fellows in CN and DBP
   b. Establishment of structured training programme for CN and DBP
   c. Conducting CME and CPD in service training for CN and DBP subspecialist and for general paediatricians and other child healthcare professionals
   d. Promotion of research and publications of local studies
5. Workshops : special workshops were held regularly for members as below
   a. Epilepsy
   b. Botox
   c. Dyslexia/ASD/ADHD
   d. EEG/Neurophysiology
   e. Augmentative and Alternative Communications (AAC)
   f. Others
6. Advocacy: the Society alerted professionals of the existence of Dyslexia in the Chinese Language and initiated the Position Statement for Dyslexia and for Attention Deficit/Hyperacidity Disorders (ADHD) which remains cornerstones for management of these disabilities. We have also advocated for the Autistic Spectrum Disorders (ASD). We work hard to provide early identification, assessment and intervention for individuals with these disorders (Dyslexia, ADHD, ASD) in the form of remediation, accommodation and compensation and urge the Hong Kong SAR Government to prioritize resources and support at home, in schools and across the community through the Equal Opportunity Ordinance and to provide special provisions at both the school and in the public examinations.
7. Parent Groups: The society works closely with parent groups for children with special needs for health education, professional guidance, advocacy, self-help and government subsidies and provisions.
8. Research: Dyslexia, ADHD, ASD, Epilepsy, AAC and others with good results and outstanding research publications.
9. Interdisciplinary collaboration with other healthcare professionals in Hong Kong via joint meetings, research projects, patient activities and others.
10. Collaboration with Professionals outside Hong Kong: Hong Kong remains one of the leading professionals in CN and DBP within the Chinese Speaking Communities and amongst pioneers for the Asia-Pacific Region.

In essence what have we achieved a lot over the past two decades and these can be summarized in the following missions accomplished:

1. Promote standard of Child Health and General Paediatrics in Hong Kong
2. Define the subspecialties of CN and DBP
3. Coordinate these subspecialties with general Paediatrics
4. Assure standard and quality of services for CN and DBP in Hong Kong.
5. Advocate for the welfare and benefit of children with special needs in the community.
6. Collaborate with other medical specialties, nurses and allied health professionals within the neurosciences.
7. Enhance rehabilitation of children with special needs.
8. Promote transdisciplinary team approach for management of children with CN and DBP.
9. Assist to set up CN and DPB services within the upcoming Children's Hospital for Hong Kong in the domains of services, training and research
10. Put the name of CN and DBP onto the map of Chinese Speaking Communities, the Asia-Pacific Region and the global world such as the International Pediatric Association (IPA) and the World Health Organization (WHO).

We are proud of our achievements and results. To all these, we would like to thank our world experts, Course Directors who conducted our Annual Scientific Meetings each year, speakers at our professional clinical conferences, fellow members of the healthcare professionals, our patient group for their support and participations. Above all, I would like to thank all the Council Members and our devoted members for their support and guidance. At the time of our 22nd Anniversary, we would like to show our heartfelt appreciation to all the pioneers (Professor C Elaine Filed, Dr. Lui Wai-ying, Dr. Flora Baber, Professor JH Hutchison and others), senior members of the Society for their contributions and to our incoming Council Members for advancing the touch of our Society forwards. We are confident that with our solidarity for CN and DBP as well as our ever-unfailing enthusiasm, the future of HKCNDP should be promising and the skyline of achievements unlimited!

I wish you all reading pleasure and best of health!

Dr. CHAN Chok Wan

Editor-in-Chief, *The Brainchild*
President, The HK Society of Child Neurology & Developmental Paediatrics
8th July 2016.

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Care of Children with Epilepsy at BC Children's Hospital

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Introduction

BC Children's Hospital is the only paediatric tertiary care center in the province of British Columbia (BC), Canada, serving a population of 4.7 million people. It is the major treatment, teaching and research facility for children in the province. The Division of Neurology and the Comprehensive Epilepsy Program provide specialised services for diagnosing, investigating and managing epilepsy which are not available elsewhere in the province. This includes care by pediatric epileptologists and other subspecialists, neurophysiology services, and specialised neuro-imaging. In children, epilepsy is one of the most common neurological condition. This article will describe the epilepsy program at BC Children's Hospital and how epilepsy care is provided by a large team from the neonatal period to young adults via various outpatient and inpatient services.

Background

What is a Seizure/Epilepsy?

Epilepsy is one of the most common neurological condition. It peaks in children under five years of age, especially the first year of life, and the elderly. Seizures are caused by excessive electrical discharges in a group of neurons. These discharges can arise from different parts of the brain, leading to various clinical manifestations, from brief lapses of attention, autonomic or psychiatric symptoms to severe and prolonged convulsions. Seizures can also vary in frequency, from less than 1 per year to many per day.

Having a seizure does not necessarily mean a child has epilepsy. Epilepsy was defined as a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures. The International League Against Epilepsy (ILAE) defines it as having at least two unprovoked or reflex seizures occurring > 24 hours apart, one unprovoked seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two, unprovoked seizures occurring over the next 10 years, or a diagnosis of an epilepsy syndrome. Epilepsy is considered to be resolved for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure-free for the last 10 years, with no seizure medications for the last 5 years[1].

What is Treatment Resistant Epilepsy?

Approximately 30 % of people affected by epilepsy are refractory to treatment.

This is often accompanied by learning disabilities, behavioural problems, memory loss, psychiatric disorders and/or other adaptive problems. It is proposed that drug resistant or medically refractory epilepsy are defined as failure after adequate trials of two tolerated and appropriately chosen and used anti-epileptic drug (AED) schedules (whether as monotherapies or in combination) to achieve sustained seizure freedom[2].

Etiology and Management

Epilepsy has numerous causes, including structural, metabolic, genetic and inflammatory. In some situations, the cause is unknown, although with advancements in neuro-imaging and genetic testing, this group is becoming smaller. The Epilepsy program at BC Children's Hospital aims to provide the most up-to-date tools to diagnose and manage patient. Management of seizure and epilepsy includes medication, surgical evaluation, ketogenic diet, vagal nerve stimulation, and providing adequate psychosocial support.

Consequences of Epilepsy short and long-term

Early diagnosis and control of seizures is crucial, as there can be many negative consequences from epilepsy. These include the effects of seizures on brain development, quality of life in the child and family, and psychosocial and mental health issues.

Uncontrolled frequent seizures are associated with an increased risk of death. Sudden unexplained death in epilepsy (SUDEP) rates are quoted to be the highest in those with treatment resistant epilepsy compared with community prevalence samples[3]. Seizures can have a negative effect on brain development and learning - especially in children. Good seizure control, even after years of treatment resistance, can have a beneficial impact on cognition[4].

With temporal lobe epilepsy, cross-sectional studies had shown that memory was worse in patients with a longer duration and earlier age at onset of epilepsy[5]. In one longitudinal study, epilepsy surgery abolished or reversed the decline in memory function[6]. In another study, 25-40% of treatment resistant patients showed decline on tests of confrontation and naming compared to friend or relative controls[7].

It also known that frequent seizures can lead to "pseudo-regression" where the seizures and medications impact sleep, energy, attention, mood, learning and interaction with the environment. This is thought to be reversible with better seizure control.

Incidence and Prevalence/Service Demand

Worldwide statistics

Approximately 50 million people worldwide have epilepsy, making it one of the most common neurologic condition. The incidence of epilepsy in children ranges from 41-187/100,000. Higher incidence is reported from underdeveloped countries. The incidence is highest in the first year of life and declines to adult levels by the end of the first decade.

The prevalence of epilepsy is higher than the incidence and ranges from 3.2-5.5/1,000 in developed countries and 3.6-44/1,000 in underdeveloped countries. Prevalence also seems highest in rural areas. This is likely due to the increased risk of endemic conditions such as malaria or neurocysticercosis, higher incidence of motor vehicle accidents, birth-related injuries, and variations in medical infrastructure, availability of preventative health programmes, and accessible care[8].

Canadian and BC statistics

0.6% of Canadians have epilepsy. Each year, an average of 15-500 people is diagnosed with epilepsy in Canada. Of this number, 44% are diagnosed before the age of 5, 55% before the age of 10, 75-85% before age 18, and 1% of children will have recurrent seizures before age 14. 1.3% are over the age of 60. This means that about 60% of new patients are young children and senior citizens[9].

In British Columbia, 40,000 individuals have epilepsy. Approximately 30% have treatment resistant epilepsy. Approximately, 3,000-5,000 patients have treatment resistant epilepsy requiring close follow-ups and continuing care. BC Children's hospital has six pediatric epileptologists. In addition, there is 1 neurophysiologist and 7 neurologists who also see patients with seizures. In the province, there is one pediatric neurologist in private practice. The remainder work at the hospital. The Division of Neurology receives over 2,000 new referrals each year and 40-50% of these are in regards to seizures or epilepsy.

Goals of the Program

Mission of Neurology Program:

To improve the neurological health of children and youth in BC through compassionate leading edge care, education and research, and to implement advances in paediatric neuroscience, particularly those that significantly improve patient outcomes.

The Epilepsy program at BC Children's Hospital is committed to providing excellent clinical management by:

- Applying advanced and up to date diagnostic and therapeutic techniques and approaches to all patients with seizures and epilepsy
- Providing holistic assessments and care for children and families
- Collaboration with other professionals to ensure a seamless service that is patient and family centered
- Providing education about the impact of epilepsy and practical management
- Participating in research to advance the field and improve patient care
- Minimizing the psychosocial impact of epilepsy for the child through education and empowerment
- Devising individual health care plans that address the needs of the child in all areas of their life, and therefore expanding their world and vision

Multi-disciplinary and Inter-disciplinary team approach

Caregivers and services involved in care

The team of physicians, nurses, healthcare professionals and support staff participate in the evaluation and treatment of our epilepsy patients who come here from across BC and sometimes other parts of Canada.

This team consists of paediatric epileptologists, neurophysiologists, paediatric neurologists, paediatric neurosurgeons, adult epileptologists, neuro-radiologists, nuclear medicine physicians, nurse specialists, pharmacologists, physiotherapists, occupational and speech therapists, dieticians, psychologists and psychiatrists, educational counsellors and social workers, and an array of scientists and technologists, who all work together to offer individualized care to children.

Program Overview

Ambulatory care/Epilepsy Clinics

A large volume of patients are seen in the outpatient setting, where the vast majority of seizure management takes place. Each epileptologist have several clinics per week. There are over 20 clinics by epileptologists each week. These consist of predominantly epilepsy patients, many whom are treatment resistant. General pediatric neurologists also see patients with epilepsy. However, if the patients have failed two anti-seizure medications or are considered treatment resistant, it is encouraged that the patients are referred to an epileptologist. Sometimes when there is a delay for pre-surgical evaluations, patients with epilepsy may suffer from numerous co-morbidities related to uncontrolled seizures. This system allows for prompt pre-surgical evaluations, access to ketogenic diet, vagal nerve stimulation, or optimizing medical management.

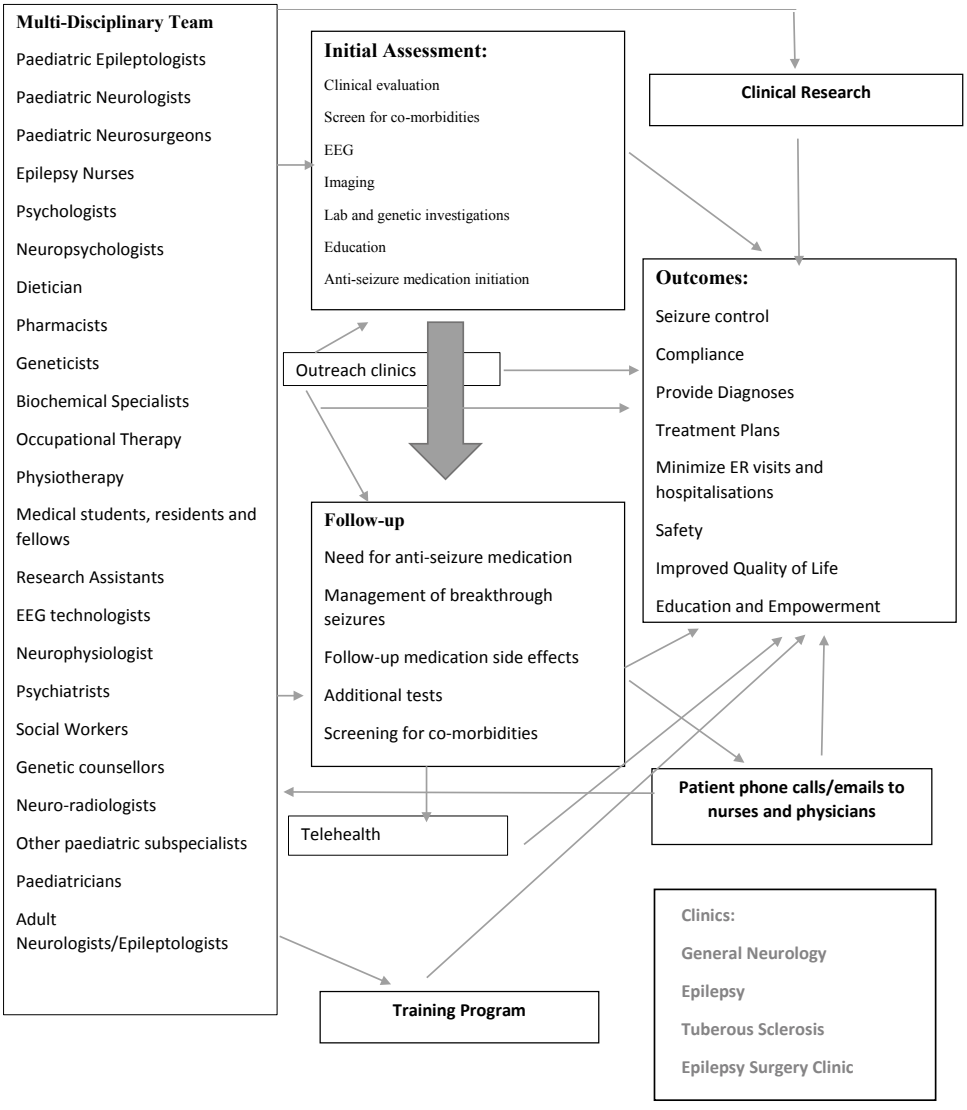
In addition to patients with epilepsy, patients with paroxysmal episodes, first time seizures, and non-epileptic events, will be seen in clinic as well. There are some specialized clinics, such as epilepsy surgery clinic, where the epileptologist and paediatric neurosurgeon meet with families together to discuss a plan for surgery or for post-surgical follow-ups. Some of the epileptologists are also involved in the Tuberous Sclerosis Clinic, where a large percentage of the children have epilepsy. Since tuberous sclerosis is a multi-systemic condition, other specialists can join the clinic or see the patients the same day. These clinics also provide an opportunity for clinical research. For example, the tuberous sclerosis clinic is currently involved in a clinical trial using a novel drug, which is an mTor inhibitor, for patients with treatment resistant epilepsy.

The care of patients in clinic involves a team of health professionals. Neurology nurses provide patient education in clinics. They are also the contact person for the patients if any concerns or issues arise. For ketogenic diet evaluations and follow-ups, the ketogenic dietician and nurses meet with the patients in clinic. The epilepsy surgery nurse is involved with teaching and support around surgery, as well as adjusting the vagal nerve stimulator.

Trainees, including medical students, pediatric and adult neurology residents, general pediatric residents and epilepsy fellows, are present. One day per week, a neuropsychologist is available to meet with patients who may be experiencing cognitive difficulties and who are not necessarily surgical candidates. Many clinical trials are also done in the clinic setting, such as drug trials, assessments of quality of life and mental health, etc. Research assistants are frequently present at clinics assisting in clinical research.

In the clinics, a thorough history and physical exam are performed. Investigations, such as laboratory testing, genetic testing, routine and ambulatory EEG's and neuro-imaging, facilitate proper diagnosis and management. Anti-epileptic medications are often initiated or adjusted. Patients are followed in clinics regularly as needed. Refer to Table 1 for a summary of ambulatory clinic care.

Table 1: Ambulatory Epilepsy Clinics at BC Children's Hospital: Improving Outcomes



Inpatient services

Neurologists on call do consultations on the inpatient wards, neonatal and paediatric intensive care, and emergency room. Patients with seizures or epilepsy that require hospital stay are usually admitted under the Neurology service. Complex epilepsy patients on the inpatient service are usually followed or co-managed by an epileptologist.

Video EEG Monitoring (Non-Invasive)

BC Children's Hospital has a two bed EEG monitoring unit. At least two patients are electively admitted every week. It is used to either confirm the diagnosis of epilepsy, or confirm the first part of a series of investigations for the possible role of epilepsy surgery for the treatment of epilepsy. Approximately 100 children are admitted per year to this unit. Patients are usually admitted for up to five days to the Epilepsy Monitoring Unit with placement of scalp EEG electrodes. The goal of video EEG monitoring is to record the patient's typical seizures. For this reason, the anti-epileptic medications are usually tapered down and frequently discontinued within the first few days of the patient's stay in hospital. The seizures are captured with video and EEG recordings and upon the completion of the evaluation the patient is reloaded on their anti-epileptic medications. The monitoring allows not only to diagnose a seizure problem accurately, but also to design the best possible treatment plan.

Investigative Tools at BC Children's Hospital

Outpatient EEG

The EEG is often used to investigate a patient's history of seizures. The "interictal EEG" describes abnormalities seen in patients between seizures. It is well known that even when patients appear well and have no outward manifestation of seizures, the brain waves can show abnormalities suggestive of a seizure tendency or risk. The "ictal EEG" describes the EEG changes that are seen during a seizure. This finding is not commonly seen during the routine outpatient EEG recordings. The capture of a seizure confirms the diagnosis of epilepsy and helps in identifying the specific epilepsy syndrome or disorder the patient has. Prolonged EEG recordings lasting several hours are often done as well. This is especially useful to characterise frequent paroxysmal events in patients.

Ambulatory EEG

Ambulatory electroencephalography monitoring is a technology that allows prolonged EEG recording in the home setting. Its ability to record continuously for 24-48 hours increases the chance of recording an ictal event or capturing interictal epileptiform discharges. This is a less expensive alternative to inpatient monitoring. It is also clinically useful, as some patients are more likely to have a paroxysmal episode in the home setting. However, medications are usually not weaned during this test for safety reasons.

Laboratory tests

Further investigations, including blood work is useful to aid in diagnosis, as well as monitoring side effects of anti-seizure medications. Common investigations include hematology profile, electrolyte panel, liver and renal function tests, drug levels, tests for inborn errors of metabolisms, autoimmune, endocrine, and vasculitic conditions. Genetic tests include karyotype and chromosomal microarray. In some cases, a lumbar puncture is needed for further metabolic, inflammatory or autoimmune work-up.

Genetic testing

In a population based study in Minnesota, approximately half of children had an unknown etiology for their epilepsy, and of the remainder, 22% were genetic and 28% were structural/metabolic[10]. The number of patients where etiology for seizures is unknown is diminishing with advances in genetic testing. In addition to karyotype or chromosomal microarray, if no etiology has been determined for seizures, further genetic studies can be done. Advances in genomic technologies, including targeted next generation sequencing and whole exome sequencing (WES), enables identification of pathogenic variants in 10–78 % of patients with unexplained epilepsy[11]. The clinical impact is significant including earlier diagnosis and specific treatment interventions.

Between December 2014 to June 2015 WES was performed in 43 patients under a study protocol. A definite diagnosis was made in 13 patients. Obtaining a diagnosis has numerous benefits, including preventing excess investigations, tailoring management options, screening patients for other co-morbidities, and providing closure to families. All patients receiving testing obtain pre and post-test genetic counselling.

Imaging

Magnetic Resonance Imaging (MRI) Scan

MRI scans have become a crucial component in the evaluation of patients with epilepsy and are particularly helpful in the evaluation for possible epilepsy surgery. Advances in technology have also helped determine the cause of seizures in patients who were called "idiopathic" in the past. The MRI shows the gross appearance of the brain and therefore can give anatomic and pathological information of the brain. The use of the MRI scan in patients with epilepsy has been aided by FLAIR techniques and High Resolution or 3T MRI imaging. High Resolution MRI with thin slices of the brain facilitates in identifying subtle lesions, such as cortical dysplasias. It is usually used in patients with focal epilepsy, in whom no lesion has been detected on the standard 1.5T MRI.

Positron Emission Tomography (PET scan)

Positron Emission Tomography (PET scan) is a neuro-imaging test of how the brain functions. PET scans can be used in the evaluation for epilepsy surgery. FDG (fluoro-deoxy-glucose) PET is the most frequently used PET scan in epilepsy patients and provides information on how various regions of the brain utilize glucose. Areas of the brain that do not

metabolize glucose may reflect the dysfunctional region of the brain giving rise to seizures. This test can be helpful in providing additional information for accurately locating epilepsy in a specific region of the brain. This is occasionally used in epilepsy surgery evaluations at BC Children's Hospital.

Ictal Single Photon Emission Computed Tomography (SPECT) Scan

Single Photon Emission Computed Tomography (SPECT scan) is another functional neuro-imaging test that helps in the localization of epilepsy to one region of the brain. SPECT scan measures the relative blood flow in various regions of the brain at a specific moment in time, i.e. when a particular pharmacologic tracer is injected. When the tracer is injected soon after the onset of a seizure, information of blood flow during a seizure can be obtained. Areas that "light up" during this test may reflect the areas of the brain in which the seizure begins. SPECT scans are usually done when the patient is admitted for video EEG monitoring; however interictal SPECT scans can be done as an outpatient.

Magnetic Resonance Spectroscopy (MRS)

Magnetic resonance spectroscopy uses techniques from MRI imaging to give quantitative chemical information of the brain. The information is usually displayed graphically, showing where there is too much or too little of certain chemicals in specific regions of the brain. Recently, a patient at BC Children's hospital had received treatment for resistant seizures. MRS led to the diagnosis of creatine deficiency, and treatment could be initiated.

fMRI

Functional MRI (fMRI) is based on increased cerebral blood flow during brain activation using blood oxygenation level dependent (BOLD) contrast. Blood flow increase exceeds the increase in local cerebral oxygen and this leads to localized increase in the ratio of oxyhemoglobin to deoxyhemoglobin. It can localize brain function and functional deficit. At BC Children's hospital, this is frequently used for pre-surgical evaluations and has replaced the WADA test for language lateralization, as it is less invasive.

Epilepsy Surgery Program

Various epilepsy surgery procedures can be used to cure or reduce seizure frequency. Most procedures are either designed to resect or disconnect the area of the brain where seizures originate or spread. Epilepsy surgery is a procedure that could either remove or isolate the area of the brain where seizures originate. Epilepsy surgery can significantly improve seizure control in carefully selected individuals. The percentage of seizure freedom can be as high as 70-80%, and a large percentage of patients can have a significant reduction in seizure frequency or fewer disabling seizures following surgery. However, referral for evaluation often is delayed and occurs years later after numerous medications have been tried. Unfortunately, it often takes 20 years for a patient to be referred for evaluation for epilepsy surgery.

Worldwide, epilepsy surgery is underutilized. Less than 1% of patients with treatment resistant epilepsy are referred for a surgical evaluation. Lack of knowledge by physicians of the benefits of surgery and appropriate surgical candidates, fear of complications, and the thought that people may outgrow the epilepsy at a later time are some of the reasons. Another reason could be due to physicians' perception of epilepsy surgery as a "last resort" procedure.

In British Columbia, 40,000 individuals have epilepsy. Approximately 30% have treatment resistant epilepsy. Therefore, 3,000-5,000 patients in our province could benefit from epilepsy surgery. The goals of our epilepsy program are to decrease seizure frequency or to render patients seizure free sooner with better success than medical therapy. In children, the aim is also to prevent the negative effects of seizure on brain development, and to improve quality of life in the child and family. Better and earlier seizure control should reduce seizure associated morbidities.

When medications are unable to control seizures, a careful evaluation is done to determine the best alternative for treatment. The goal of the pre-surgical evaluation is to determine the epileptogenic zone: the part of the brain that is integral for the generation of seizures, and to determine if this can be safely resected. This work-up involves a detailed patient history, description of seizure semiology, video-EEG monitoring to localize seizures, neuro-imaging to identify the type, location, and extent of structural abnormalities, and nuclear medicine tests, such as SPECT and PET scans for seizure onset localisation. Assessments of cognitive functioning, including neuropsychological evaluations and fMRI can be used to assess cognitive co-morbidities associated with the epilepsy and predict the cognitive outcome of patients after surgery. The work-up and investigations are tailored to each patient. In some patients, the seizure focus cannot be fully identified and further invasive monitoring is necessarily. On average 18-20 children have epilepsy surgery each year in the program. With an increasing number of new epileptologists, the number may change. Please refer to Table 2 for a summary of the pre-surgical evaluation at BC Children's Hospital.

Invasive Monitoring

Invasive monitoring is used in pre-surgical evaluations at BC Children's Hospital if the epileptogenic zone cannot be localised by surface EEG and other investigations. Invasive EEG recordings are made with surgically implanted electrodes on the surface or within the depth of the brain. Subdural EEG electrodes are those electrodes which are placed over the surface of the brain. Depth EEG electrodes are those electrodes which are placed within the parenchyma of the brain. These recordings are done to better localise the region of the brain from which the epilepsy is arising. The placement of these electrodes is often confirmed with co-registration on an MRI scan image.

Functional Localisation Techniques

Functional localisation techniques are performed to predict or prevent functional deficit with epilepsy surgery. These are done separately during the operation or prior to the operation by the epileptologists and EEG technologists.

Cortical stimulation entails delivering a short duration of electrical current to the brain via the invasive electrodes. This is done to determine what function that region of the brain subserves. Patients are often asked to perform a task during cortical stimulation, such as speaking, to determine if stimulation impairs the performance of that task. Functions that can be tested using this technique include language, vision, movement, and sensation.

Cortical somatosensory evoked potentials are used to locate an important structure in the brain, called the central sulcus, which helps to delineate motor from sensory function.

Neuropsychological Evaluation

Research has shown that many people with epilepsy have cognitive difficulties as a result of their seizures. The type of cognitive difficulties people experience depends on the area of the brain the seizures are coming from. For example, those with frontal lobe epilepsy can have difficulties with executive function and attention. Patients with left temporal lobe epilepsy may have difficulties with short term memory and verbal memory. Many people with epilepsy also experience depression and anxiety that can affect their thinking skills. A neuropsychological evaluation is conducted by a neuropsychologist as a formal assessment of cognitive abilities (e.g. memory, concentration, and problem solving), mood and personality. Pre-surgical patients have priority for testing. Patients are periodically followed post-operatively as well.

Vagal Nerve Stimulator (VNS)

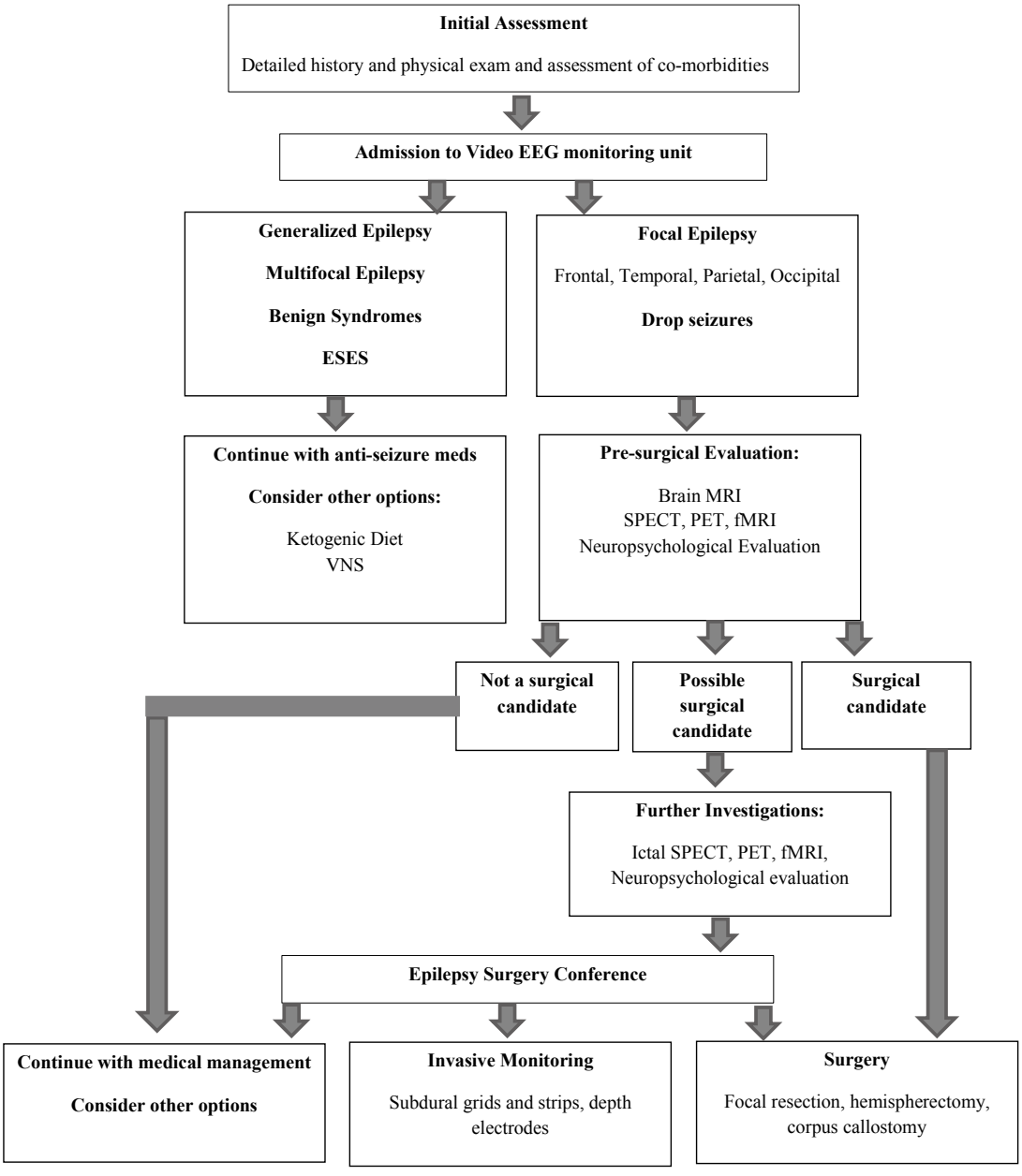
At BC Children's Hospital, VNS is used in those with treatment resistant epilepsy, who are not epilepsy surgery candidates or who cannot tolerate the ketogenic diet. It is a palliative therapeutic modality with a response rate of 50% reduction in seizure frequency in one third to one half of patients. It delivers electrical stimulation to the left vagus nerve. The patients are followed in clinic and the epileptologist or epilepsy surgery nurse adjust the settings periodically for optimal response. From 2006, 57 VNS's have been implanted, with approximately 3-5 devices being implanted per year. There are currently approximately 30 patients being followed with VNS in the program. The remainder have transitioned to adult care, passed away or have had the device stopped.

Epilepsy surgery conference:

After the necessary tests are completed, cases are presented at Epilepsy Surgery Conference. This conference occurs for 2 hours every second week. Here, a multidisciplinary group — including epileptologists, a neurophysiologist, neurosurgeons, neuropsychologists, neuroradiologists, nurses, and trainees, gather to review and discuss all the data collected. If

the data clearly shows a definite, localised seizure focus that can be removed without creating significant loss of function, the decision to offer surgery can be made at this point. If not, further investigations or treatment options are suggested by the group. Other professionals may be consulted to further assistance with the surgical experience.

TABLE 2: EPILEPSY SURGERY EVALUATION



Ketogenic Diet Program

The Ketogenic Diet (KD) is a strictly calculated, medically prescribed therapeutic diet that has proven efficacy for treatment of intractable childhood epilepsy. The classic KD (also known as the long-chain triglyceride diet) is a high fat, low carbohydrate dietary therapy.

Although double-blind control trials are lacking, there is sufficient evidence to suggest that the classic KD if effective in reducing seizure frequency (>90% reduction) in approximately 30% of patient with treatment resistant epilepsy. If effective and tolerated, children remain on the KD for 2 to 3 years, at which point it is often weaned and discontinued.

The ketogenic diet team at BC Children's hospital consists of 1 dietician, 2 nurses, and 3 epileptologists. If another neurologist or epileptologist considers starting a patient on KD, they will refer the patient to this team. In the program, the diet is introduced in the outpatient setting over four days for medically stable patients over one year of age. Rarely, very young patients or complex patients initiate the diet as an inpatient on the wards or ICU. Comparisons of inpatient and outpatient treatments using a KD have been done. It has been shown that the benefits of outpatient treatment include improved acceptability and ability to maintain and comply with the diet. It also avoids the expense, inconvenience and potential low blood sugar associated with starvation during inpatient initiation[12]. The initiation of the diet varies based on whether the patient is on oral diet versus formula diet and whether or not they are older or younger than one year of age. In some situations, the ratio is progressed (with full calories) but in some cases the calories are progressed (at goal ratio). The program currently has 40 patients on the diet. On average, 1 – 2 patients are initiated on the diet per month. Recently, the patients on the diet are asked to complete forms on quality of life and psychosocial functioning to assess the impact of the diet on other aspects of their lives, besides seizure frequency.

Telehealth and Outreach Clinics

Telehealth

The epilepsy program is based primarily at BC Children's Hospital. Access to epilepsy clinics can be difficult for all children in the province. Geographically, British Columbia is a large province and many patients live at a great distance from the hospital or in remote areas. There is also disparity with access to transportation and socio-economic status. The epileptologists use telehealth for some follow-up appointments. Assessment and care planning is done over a videoconferencing system rather than traveling to access the same services. All telehealth services are provided over a secure network so that privacy is ensured. More than 40 BC communities have accessed to the services.

Outreach Clinics

Some of the neurologists and epileptologists also go to different areas of the province to see patients who live in distant or remote areas from BC Children's Hospital. These include 5 different sites in the province. From 2014-2015, there were 718 outreach clinic visits, of which approximately 40-50% of these were seizure or epilepsy patients.

Quality of Life

In addition to seizures, patients with epilepsy have a higher risk of emotional, behavioural, social, cognitive, and academic and family problems when compared to healthy peers and children with other chronic health conditions. These can affect a child's quality of life, including physical, social and mental well-being, and functioning in daily life. When treating patients with epilepsy, it is important to monitor the degree to which epilepsy affects their life. Assessments of quality of life can document the patient's status, including how their status is before and after treatment changes.

Recently, the epilepsy group starting using the Pediatric Quality of Life Inventory to patients for monitoring this. These questionnaires are filled out by the patient or parent, depending on the child's age. Patients who are given this questionnaire include those who are newly diagnosed with seizures, patients in the epilepsy surgery program, and those who are on the ketogenic diet. In addition, the ADHD-IV Rating Scale, Strengths and Difficulties Questionnaire, and Parental Stress Scale are administered. If there are any concerns, referrals can be promptly made to psychology or psychiatry.

Transition Planning to Adult Care

Significant improvement in medical management over the last decades has resulted in more children surviving to adulthood. For adolescents with epilepsy there is very little information on the best way to transition. At BC Children's Hospital, patients are seen up to 18 years of age. Usually, planning for transition begins 1-2 years earlier. A detailed referral summarizing the patient's history and investigations is sent to the adult physician. Patients with treatment resistant epilepsy, complex patients, those who require epilepsy surgery work-up or have had vagal nerve stimulation, are referred to adult epileptologists. The remainder are referred to adult neurologists in the province.

At BC Children's Hospital, a group of physicians including 5 neurologists (1 adult and 4 pediatric), adolescent medicine specialists, family practitioners, psychiatrists, and 7 allied health professionals, created long term care plans to ensure access and attachment to adult healthcare providers and to identify strategies for providing support during the transition period including training modules, tools, and resources and system improvements. Different aspects of epilepsy have been covered, such as patients with treatment resistant epilepsy, epilepsy surgery patients, women in epilepsy, etc. The transition development is still in progress. However, the process has so far resulted in a collaborative approach between paediatric and adult healthcare providers. It has helped to create plans to better organise and optimize the transition process and has provided a forum to identify and remedy gaps in the transitional care process.

Patient Education

The epilepsy program aims to provide seizure management and safety information to patients and families, as well as to minimize the psychosocial impact of epilepsy for the child through education and empowerment.

In addition to the physicians, the epilepsy nurses play an important role in patient education. They help educate the child, family, respite services, etc. about the full impact of epilepsy. Epilepsy nurses also help with practical management, including emergency medication, seizure first aid, and Sudden Unexplained Death in Epilepsy (SUDEP). They build and sustain long term relationships with the child and family and ensure they understand the information provided by the physician. They also provide practical advice regarding medication administration. They are an accessible point of contact for the child and family and address any issues arising. Handouts and brochures are available to families on various topics, including descriptions of epilepsy, epilepsy safety, and anti-seizure medications.

The BC Epilepsy Society is a provincially incorporated non-profit charitable organisation, dedicated to serving people in British Columbia with epilepsy and their families. The organization provides support and education for patients and families in various ways, such as providing a resource center, lecture series for families, and educational events. They also provide hospital outreach, where staff members from BC Epilepsy Society meet with families attending the Neurology Clinic at BC Children's Hospital to provide epilepsy resources and assistance with community services. Resources are also provided to other hospitals and health clinics across the province.

Training Program

An important part of the epilepsy program at BC Children's Hospital is training medical students, neurology residents, and clinical neurophysiology and epilepsy fellows. The high volume clinical practice and varied areas of expertise of the faculty provides a rich learning environment. Trainees get exposure to a wide variety of cases in multiple clinical settings, including outpatient clinics, OR, inpatient monitoring unit, and ICU. Various academic rounds are in place, such as Video EEG session, Epilepsy Surgery conference, Epilepsy Genetics Rounds, and Epilepsy/neurophysiology lectures.

Trainees have the opportunity of attending epilepsy clinics, including epilepsy surgery clinic and tuberous sclerosis clinic. Neurology residents do a mandatory epilepsy and neurophysiology rotation in their residency.

Epilepsy and Neurophysiology fellows receive 1-2 year positions, during which they acquire extensive experience in electro-diagnostic testing, including electroencephalograms, long term video EEG monitoring, evoked potentials, and intraoperative monitoring. They become competent with clinical management, including epilepsy surgery evaluations, and following patients on the ketogenic diet. They are expected to be involved in research as well. The program has graduated several alumni who now are clinicians and scientists in many centers in the world.

Research

Research into the causes and treatment of epilepsy is very important part of the epilepsy program. The BC Children's Epilepsy Program provides an environment where basic science studies that advance the scientific knowledge of epilepsy can be translated to clinical studies that benefit patients and their families. All of the epileptologists and a large number

of students, fellows, and other staff are involved in various prospective and retrospective studies, including basic science and clinical research. At the present time, there are 11 ongoing prospective trials in the epilepsy program. Some of them involve ketogenic diet, quality of life, tuberous sclerosis complex, telehealth, epilepsy surgery, epilepsy genomics, fMRI, electrical status epilepticus in slow wave sleep, etc. Numerous retrospective studies are also ongoing. This is facilitated by a clinical epilepsy database and an extensive EEG database.

Conclusion

The high-volume clinical practice at BC Children's Hospital provides care to paediatric patients from all over the province of British Columbia, as well as occasionally other provinces in Canada. A coordinated, multidisciplinary team contributes a comprehensive range of skills and knowledge of paediatric epilepsy. Excellence in diagnostics, medical and surgical clinical treatment programs, and research have yielded important contributions. There are still areas that are work in progress, such as improving the transition process to adult care, having more specialised clinics, such as neurogenetic or ketogenic diet clinic, and being able to obtain funding for specialized genetic testing or imaging modalities for patients, outside of research protocols.

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BC Children's Hospital Brain Mapping Program

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Scientific Director, Child & Family Research Imaging Facility

BC Children's Hospital (BCCH), the primary paediatric teaching hospital of the Faculty of Medicine of the University of British Columbia in Vancouver, provides pediatric neurological and neurosurgical care services to the province of British Columbia, and is a major Canadian academic paediatric neurosciences centre, contributing tertiary and quaternary care services to children with a wide range of neurological disorders. Children with neurological disorders often require several different consultations and diagnostic tests to reach a precise diagnosis and to plan an optimal treatment strategy to achieve the best outcome, often requiring a team approach to care. Comprehensive multidisciplinary team evaluation is especially relevant to children with epilepsy, brain tumors and strokes, who may need to undergo medical and neurosurgical intervention as part of their care. The BCCH Brain Mapping program was created to enhance neurological evaluation and surgical planning by teams of paediatric neurologists and neurosurgeons.

The BCCH Brain Mapping program provides individualised surgical planning guidance by combining structural neuroimaging, to locate brain pathology to be targeted by neurosurgeons, with *functional neuroimaging*, to highlight areas responsible for important brain functions, which are in close proximity to surgical targets, in order to minimize damage to those areas. Brain functions of particular importance include motor function (especially hand fine motor function), vision, hearing, speech and language.

In developing the BCCH Brain Mapping program, our initial key goals were: (1) to reduce reliance on previously available invasive methods of mapping of brain function (unilateral carotid artery injection of amobarbital and awake brain surgery with direct cortical electrical stimulation); (2) to avoid exposure to ionizing radiation (CT, PET); (3) to design a functional neuroimaging laboratory environment that was child-friendly; (4) to design tasks adaptable to a variety of levels of difficulty and to measure behavioral performance, cerebral neurophysiology and brain anatomy simultaneously; and (5) to visualize and present multimodality, multidimensional brain data in a format that would help guide pediatric neurological and neurosurgical evaluation and care.

To achieve these goals, over the past twenty years, we have created infrastructure to specifically develop and support paediatric brain mapping technology and methodology with BC Children's Hospital, and we have progressively translated advances in technology into advances in neurological care of children.

A. Motivation, Challenges & Opportunities

What motivated us to develop a paediatric brain mapping program at BCCH? The main motivation was the opportunity to enhance paediatric neurological care by applying advances in MRI and computing technology to enable fusion of three-dimensional images of changes in brain physiology (correlated with behavioural events), with three-dimensional images of brain anatomy, to achieve interactive *in vivo* spatiotemporal mapping of brain function. This presents many associated challenges. Most fundamental of these is the fact that the brain is an incredibly complex organ consisting of nearly 100 billion (10^{11}) neurons, with approximately one million billion (10^{15}) connections among them. Connections form during early fetal life under the influence of genetic and environmental factors, with ongoing development and refinement occurring throughout infancy, childhood and adulthood. All the diversity and complexity of human perception, cognition, thought, emotions, memories and behaviour result from interactions among neurons participating in multiple, dynamic, distributed brain networks. These, in turn, are organised according by location and connections in the brain (neuroanatomy and connectivity), and governed by constraints of neurophysiology (e.g., metabolism, electrical transmission, neurotransmitters and receptors).

In the past, in order to determine which areas of the brain are most responsible for critical functions, neurological deficits observed in life were correlated with results of examination of the brain after death (clinical-pathological correlation). Subsequently, it became possible to observe the effects of temporary disruption of brain activity on living persons by direct electrical stimulation of the brain during awake brain surgery, a method pioneered by neurosurgeon Dr. Wilder Penfield in Montreal. Another method of temporary perturbation of brain function, described by Dr. Juhn Wada, involved selective unilateral injection of the anesthetic agent, amobarbital, into either the left or the right internal carotid artery during awake cerebral angiography, and then observing for deficits of language or memory, to determine whether one hemisphere appeared to be "dominant" for language or memory. We were motivated by the opportunity to provide similar information without reliance on such invasive methods.

As more powerful superconducting magnets became available for MRI, higher-resolution structural images became possible, with better delineation of tissue boundaries making possible more accurate quantitative measurement of grey and white matter volumes in areas of the brain, during development (brain image segmentation, parcellation and morphometric analysis).

Meanwhile, MRI studies have demonstrated that deoxyhemoglobin (paramagnetic, or weakly magnetic, due to unbound iron in heme) and oxyhemoglobin (diamagnetic, with minimal magnetic effect) have different effects upon the MRI signal in various areas of the brain, and that these effects vary, moment by moment, with transient changes in neuronal activation. Thus, areas of the brain, that are receiving incoming neuronal stimulation ("activation"), experience a temporary increase in the locally measured MRI signal due to

transient local increases in blood oxygen content. This phenomenon is termed the "Blood Oxygen Level Dependent (BOLD)" effect, and is the basis of "functional MRI".

Other MRI studies explored the diffusion of water within various types of tissue (diffusion weighted imaging), leading to the observation that diffusion of water was favoured in the direction of axons within white matter tracts of the brain. This led to the development of MRI techniques for "diffusion tensor imaging (DTI)" to demonstrate white matter pathways ("tractography").

High-resolution anatomical (structural) MRI can now be combined with studies of structural (white matter tractography) connectivity and BOLD functional MRI connectivity, acquired either during specific tasks or at rest (intrinsically autocorrelated functional networks), and such studies can be performed non-invasively on infants, children and adults.

B. Creating a Paediatric Brain Mapping Team

The initial concept of a Paediatric Brain Mapping program at BCCH was the result of a careful long-term planning by a small "Brain Mapping Steering Group" (Drs. B. Bjornson and P. Wong , paediatric neurologists; Dr. D. Cochrane, paediatric neurosurgeon; and Dr. D. Giaschi, vision scientist), subsequently enhanced through collaboration with others (particularly Dr. A. Mackay, MRI Physicist; and Dr. S. Miller, neonatal neurologist). Progress has depended upon the talents and innovations contributed by many individuals, including undergraduate and graduate students in several disciplines (psychology, physics, engineering, computer sciences, medicine and neurosciences). Our current Brain Mapping & Neurotechnology Laboratory team consists of a Brain Mapping Technology Development Manager, with expertise in experimental design and neuroimaging; a Research Imaging Technology Integration Manager, responsible for integrating EEG, MEG and other physiological data with functional MRI data; and a Brain Mapping Neuroinformatics Engineer, with expertise in software development and computing. We currently collaborate closely with several MRI Scientists who optimize MRI pulse sequence software and MRI hardware performance to enhance MRI data quality and yield, and with Neurologists, Neurophysiologists, Neuroscientists and Computer Scientists with additional expertise in digital EEG, experimental design, image processing and statistical analysis.

C. Customizing Brain Mapping Task Protocols for Children

We have developed customized paediatric experimental protocols and software, which allow us to select functional MRI protocols suitable for children of multiple different levels of ability. We have nearly 100 different versions and levels that can be used to map visual, auditory, language and visuospatial functions in children.

D. Creating a Child-Friendly Brain Mapping & Functional MRI Lab Environment

Our brain mapping team designed a child-friendly MRI simulator unit (see Figure 1), in which we are able to acclimate children to the environment of the MRI scanner, and familiarise them with testing protocols, to increase success rates by allaying anxiety, enhancing understanding and cooperation, and ensure that procedures are matched to each child's level of ability. We have designed a matching facade for the GE Discovery 750 3.0 Tesla MRI system (see Figure 2) that we currently use for paediatric functional MRI.

Figure 1. The Child-Friendly Paediatric MRI Simulator Unit in the Brain Mapping and Neurotechnology Laboratory at BCCH.

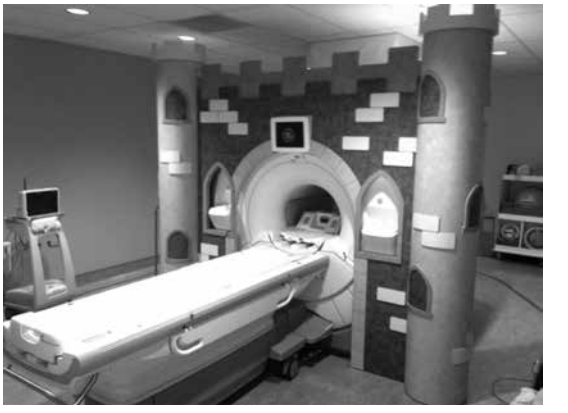

Figure 2. The Child-Friendly Exterior Façade of the GE Discovery 750 3.0 Tesla MRI scanner located in the Child & Family Research Imaging Facility at BCCH.

E. Presenting Results to Paediatric Neurologists & Neurosurgeons

We have designed a consistent image presentation format for showing results of functional MRI studies of children, using panels of axial, coronal and sagittal images, as well as multi-planar reformatted images with concentric erosion to demonstrate activation at depths of cerebral sulci. We prepare and present three-dimensional renderings images in orientations simulating the position in which a child will be oriented during neurosurgical procedures.

F. Impact on Paediatric Neurological and Neurosurgical Care

We have scanned over 150 children who were referred for neurosurgical planning, most of whom were candidates for paediatric epilepsy surgery. As a result of our experience, expertise and publications, paediatric neurologists and neurosurgeons at BCCH have stopped requesting unilateral carotid artery amobarbital injections for determination of cerebral language dominance. Neurologists and neurosurgeons rely upon the data that are presented for clinical decision-making. We are currently expanding our capabilities to include simultaneous acquisition of 3T pediatric functional MRI and 256channel EEG.

G. Impact upon Pediatric Brain Mapping & Functional MRI Infrastructure and Research at BCCH & UBC

In achieving these goals, our Brain Mapping and Neurotechnology Laboratory team became the core technology support group for a new dedicated paediatric 3T MRI research scanner, funded by a federal grant from the Canada Foundation for Innovation, and housed in the new BCCH "Child & Family Research Imaging Facility", one of only two dedicated paediatric 3T MRI research facilities in Canada (see Figure 3).

Figure 3. The Child & Family Research Imaging Facility at BCCH.

H. Collaborative research publications involving the BCCH Brain Mapping & Neurotechnology Laboratory

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Diagnostic Neurophysiology Services

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Clinical Services

We provide neurodiagnostic testing for all paediatric and maternity patients at Children's and Women's Health Centre of BC (190 beds), and we also accept referrals from paediatricians and general practitioners in the community. We are the only dedicated paediatric and maternity hospital in the province of British Columbia (population 3.5 million). Because of our uniqueness, we see many interesting and difficult patients. This means that our technologists require a high level of expertise and need to stay on the cutting edge of techniques and technology.

The department is under the direction of Dr. P.K.H. Wong and includes five electroencephalographers, one electromyographer, two Epilepsy Fellows, 13 technologists, one biomed data coordinator and four clerical support personnel.

Our service is unique in that all modalities of neurophysiological diagnostic testing are provided in one department. We do this by performing 26 different procedures on all levels of the nervous system. For example, procedures include EEG, cEEG, EMG, Evoked Potentials of all modalities, and Intensive and Ambulatory EEG Monitoring. Our Epilepsy surgery work-up includes electrocorticography, motor strip and speech localisation, and indwelling subdural recordings. We also offer intraoperative monitoring for Orthopaedic and Neurosurgical cases. This includes somatosensory and motor evoked potentials, cranial nerve monitoring, deep white matter stimulation and free-running and triggered EMG during lipoma removal and selective posterior rhizotomies.

This document gives an overview of how each service is provided.

EEG

Children are sleep deprived to obtain a natural sleep recording where possible. All EEGs are performed with simultaneous video, and parents accompany their children during the test. For safety and accuracy of electrode application, patients who have a tendency to move a lot are secured into a papoose board. A Velcro strap is secured gently across the stomach and a low tumble form roll may be placed under the neck. Parents often lie on the bed beside their child. Once the head is measured, reusable gold electrodes are applied with Ten.20 conductive paste and the hair is overlapped to hold each electrode in place. For young and uncooperative children a hat is made from Surgifix to contain the electrode wires. This more easily allows the parents to hold their infant or toddler child during testing.

Hyperventilation

Hyperventilation (HV) is performed for 3 minutes on an anterior to posterior bipolar montage with 1-minute pre- and post-HV baselines recorded. Younger children are given windmill toys to blow into. The room lights are turned up to optimise the video recording during this time.

Children presenting with questionable staring spells are asked to hyperventilate for 4 minutes. If the first period of HV shows spike-and-wave discharges, or if the patient does not fall asleep during the EEG, a second 4-minute HV is done. Children who have absence seizures in the first HV or who have treated absence seizures are asked to perform two periods of HV for 4 minutes each. Optimally, the second HV is performed with the patient sitting up in order to demonstrate any loss of body tone. Two 4 minute periods of HV are repeated when these children return for follow-up EEGs after treatment. Often, breakthrough absence seizures will be seen only during the second HV in patients on treatment.

Photic Stimulation

Flash stimulation is performed on all patients over 3 months of age recording on a bipolar montage with anterior-posterior and posterior halo derivations. Photoparoxysmal responses are most prominent on arousal from sleep. When possible, photic stimulation is done after the patient is aroused from the sleep recording.

Our protocol calls for the strobe light (0.72 joules) to be positioned 30 cm in front of the eyes. Room lights are set to minimum. Stimulation begins at 16 Hz on eye closure followed by stimulation at 1, 3, 6 and 9 Hz for a duration of 5 seconds each with an inter-stimulus interval of 5 seconds. Patients are instructed to keep their eyes closed until 9 Hz after which they are instructed to close their eyes when the light begins to flash and open them when the flashing stops. Frequency increments of 3 Hz are used, up to 21 Hz. For children younger than 2 years, stimulation is done at lower frequencies (0.5-10 Hz) while awake and with eyes open, if possible.

When a photoparoxysmal response is evoked, the strobe light is stopped as soon as possible to determine if the discharge outlasts the stimulus. Any clinical accompaniments are noted. For generalized discharges, response testing of the patient is done during stimulation.

Capturing Events and Response Testing

When the EEG referral questions whether or not a particular behaviour is a seizure, the recording is often prolonged to capture the event. EMG electrodes are applied if limb movements are involved, the room lights are turned up and the video camera image may be customized to maximize recording of the event.

Response testing can take many forms depending on the age and cooperation of the patient. Practice trials are done to form a baseline of the response time. The technologist will

instruct patients to repeat words or numbers said to them. If patients do not repeat words given to them during the discharge, they are asked to recall those words afterward. Stimuli should be presented as soon as possible after onset of the EEG discharge. If no impairment in consciousness is demonstrated, patient is asked to perform a motor activity before or during stimulation (clapping or pointing to the door). If no impairment is noted, the technologist asks quick math questions during stimulation (for example, what is 2+2?) or to state the opposite of a word given (for example the opposite of day is night). Often, patients who are able to recall words or do motor activities during a discharge are not able to perform these higher functions.

Epilepsy Monitoring Unit and Ambulatory EEG Monitoring

The demand for 1-5 days in-patient or overnight out-patient EEG recordings has been increasing. We have two dedicated epilepsy monitoring beds on the ward (with a planned increase to 6 beds in the near future) and two ambulatory EEG systems. We apply disposable disc electrodes with Collodion glue for these overnight recordings. To reduce the risk of accidental strangulation, the wires are slipped through narrow Surgifix gauze and the headbox is placed in a pouch with a belt that goes around the patient's waist. The backpack must be worn by the patient at all times during the monitoring. The parents are given instructions on response testing and rules to ensure optimal recording conditions (keeping the child in the camera range, keeping blinds closed, not chewing gum, etc.) Parents must stay with their child for the duration of the monitoring to identify events, press the event button, describe the events and perform response testing. This requires detailed instruction to parents.

24-hour Continuous EEG Monitoring (cEEG)

Continuous EEG monitoring is currently performed for comatose patients having seizures or for patients in status epilepticus. There is a great demand to expand this service to be the standard of care for all patients at risk for seizures in the Intensive Care Unit (ICU), including patients who are post-cardiac surgery and those with meningitis, encephalitis or decreased levels of consciousness.

For routine portable EEGs in the ICU, a full set of disposable disc electrodes is applied with Ten.20 cream and the hair is overlapped to hold each electrode in place. Continuous EEG recordings are often a continuation of a routine recording during which seizures are captured. To further secure the electrodes for continuous recording, Grass EC2 cream is placed on top of each electrode followed by a gauze square. A Surgifix hat is placed over the head and covers the electrode wires connected to the headbox. The headbox is secured to the head of the bed. An event button is made available for staff to press for clinical seizures. The video angle is set very wide to include staff movement around the bed. The bedside computer is plugged into a network outlet and EEG reader stations in the department are enabled for remote viewing.

These recordings are interpreted a minimum of twice a day and reported by EEG Fellows under the supervision of staff electroencephalographers. Written communication of the results occurs daily with more frequent verbal communication of results to attending physicians as dictated by the clinical scenario.

Sedated Procedures

Chloral hydrate sedation is used for EEG testing on severely uncooperative or autistic patients and for children having nerve conduction, EMG, BAER and SEP testing. These patients are sleep deprived and kept nil by mouth for 2-6 hours before sedation (depending on whether they are out-patients or in-patients). The standard dose of chloral hydrate is 40-50 mg/kg, given one hour before the test.

Nerve Conduction and Electromyography (NC/EMG)

Neuromuscular testing is most often requested for inflammatory demyelinating neuropathies, focal mononeuropathies, investigation of infants with abnormal tone and, rarely, myotonic dystrophies.

Chloral hydrate sedation is administered by a registered nurse one hour before the testing. The nurse explains the test and shows the parent and patient pictures of the procedure. Two technologists are assigned for each paediatric NC study to distract the patient and obtain accurate results as quickly as possible. One technologist runs the equipment and the other performs the study. Parents are given the option to come in with their child or to stay in the waiting area during the testing.

Our standard protocol for NC testing on the upper limb is to perform sensory and motor recordings (with F wave) on the median and ulnar nerves. On the lower limb, the following recordings are done: sural sensory and tibial and peroneal motor (with F wave). Additional nerves are tested as clinically indicated. Interpretation of results is primarily based on conduction values and the amplitude of the responses. Distances for stimulation and recording are not always a standard length due to small limb size in young patients. Rarely, repetitive nerve studies (facial nerve) are performed, but these are technically challenging in children.

A nerve conduction study typically takes 20 minutes. The electromyographer then reviews the results and if indicated, proceeds to do needle EMG studies. Attempts are made to test distal and proximal lower limb muscles (tibialis anterior, medial gastrocnemius) and one upper limb muscle (biceps). The area is often sprayed with a topical anaesthetic and a disposable needle electrode is inserted into the belly of the muscle by the electromyographer. Activity upon insertion, rest and flexion of the muscle is recorded. Additional muscles are tested as clinically indicated. For investigation of diffuse processes, testing is usually performed on the right side and the left side is left untouched for biopsy purposes.

Evoked Potentials

Visual (VEP), brainstem (BAER), and somatosensory evoked potentials (SSEP) are performed on out-patients and in-patients. Referrals are most often for biochemical or demyelinating diseases and neurosurgical tumours.

VEP

Occipital electrodes are placed according to the Queen's Square measuring system and are referred to a mid-frontal electrode. For testing of optic nerve function in children who are able to follow instructions to fixate on the screen, a pattern stimulus is used. Visual acuity is noted or tested. The patient is positioned 100 cm from the screen and is asked to fixate on a spot marked in the centre of the screen. A checker board pattern of 32x32 squares is used. For those with poor visual acuity, a larger pattern check is used. Monocular stimulation is performed. Typically 200 samples are averaged. A minimum of two trials are performed with each eye, and the latency and voltage of the P100 response are recorded. Normal values are a mid-occipital latency of <115 msec with an inter-eye variation of <8 msec.

For very young children or those who are unable to cooperate for pattern stimulation, goggle flash stimulation is used. The goggles are worn by the patient and monocular stimulation is performed to test for optic nerve dysfunction. Bilateral stimulation is performed for suspected cortical visual impairment or other diffuse disease processes. Typically 100 samples are averaged and two trials are done.

BAER

This testing is done during natural or sedated sleep when possible. Ear electrodes are referred to a vertex electrode. Tiptrode stimulating electrodes are placed gently into the ear canal after visually ensuring that there is no wax in the ear canal. Unilateral alternating 70 dB HL clicks are delivered at frequency of 10.1 Hz to each ear (with 40 dB masking to alternate ear). One thousand responses are averaged. A minimum of two trials is performed on each ear. The peak and inter-peak latency of waves I-V are recorded. If a poor response is obtained rarefraction clicks maybe tried to enhance wave I, or condensation clicks to enhance wave V.

SSEP

Children receive chloral hydrate sedation for somatosensory EPs. Upper limb stimulation is performed via the median nerve at the wrist. Peripheral responses are recorded at Erb's point, subcortical responses at C7S (N13 P14) and cortical responses from the central and midline scalp areas (N20/P22). Lower limb stimulation is performed via the posterior tibial nerve at the ankle. Peripheral responses are recorded at the popliteal fossa, followed by spine responses at L1 and T6 levels (N14), subcortical responses at the neck (N20) and cortical responses from the midline and central areas (P37/N45). Typically 500 responses are averaged. A minimum of two trials are performed and the latency and amplitude of the peak waveforms are measured.

Intraoperative Monitoring

We have a very active intraoperative monitoring (IOM) program providing daily services to orthopedic and neurosurgical departments. Monitoring is performed on scoliosis patients and neurosurgical posterior fossa tumour, lipoma and spinal cord tumour patients. Intraoperative monitoring has increased from being 20% to 35% of our total workload. Other departments are requesting intra-operative monitoring services but we are unable to provide services at this time. Recruitment to add a full time IOM Specialist with PhD credentials to our team is ongoing.

Scoliosis

Each scoliosis case is monitored by two technologists (at least one of whom is certified in Neurophysiological Intraoperative Monitoring) working closely with three surgeons who are very knowledgeable about monitoring. Motor and sensory pathways are monitored. All electrodes are applied in the Operating Room once the child is anaesthetised. For motor stimulation, corkscrew electrodes are inserted into the skull over the pre-Rolandic region (1 cm forward to C1, C3, C2 & C4). Corkscrew electrodes are also used to record the post-Rolandic sensory, midline and subcortical potentials (C3', Cz', C4', Fz, Pz, Oz and high cervical spine). Needle recording electrodes are inserted into the abductor pollicus brevis, brachioradialis, abductor hallucis and tibialis anterior muscles to record motor evoked potentials. Surface sticky electrodes are used for ulnar and posterior tibial somatosensory stimulating and peripheral recording.

Once the electrodes are applied and the patient is positioned prone, the technologists plug in the electrodes and stimulators, check the impedances and obtain pre-drape baseline trials. Stimulus parameters for SSEPs start at 15mA for the ulnar nerve and 40mA for the tibial nerve. Transcranial motor stimulation begins with single pulses at 150v, ISI 1.1, train of 5 and duration 50 usec. The stimulus voltage is increased at small intervals until reproducible responses are seen from all muscles or a maximum of 800v is reached. If responses are variable, the voltage is reduced and double train stimulation is performed. Variations in the ISI, increasing the pulse duration and varying the order of stimulation (R-L, L-R-L) are also done to optimize responses both when getting baselines or during the case.

When traction is used for scoliosis, the baseline is set before traction is applied and trials are taken for a minimum of 20 minutes before the surgeon starts cautery to open the surgical site. Limited trials can be done during cautery. Our protocol requires stimulation for transcranial motor electrical potentials (MEP) every 5 to 7minutes, and technologists will ask for permission to interrupt cautery to get a trial when possible.

Our alarm criteria for MEPs is a drop in amplitude to <50% of the baseline. For SEPs, it is a drop in amplitude to <50% of the baseline plus a latency increase of 2 msec. Our orthopaedic surgeons will not operate on the spine without monitoring. Confidence in our monitoring has enabled them to be more aggressive with corrections and has allowed

correction of more severe curves. During 35-50% of our cases, alarms have been called and interventions have been performed (reduction or release of traction, adjustment to anesthesia, increase in median arterial pressure (MAP)). Decreased amplitude and loss of responses associated with low MAP has encouraged anesthesiologists to keep the MAP above 55. Early detection of other genuine response losses has resulted in the neuro-protective protocol being started immediately when indicated.

Neurosurgical

An IOM Specialist attends most neurosurgical cases and performs the monitoring with relief provided by technologists. The parameters monitored are customised to the clinical case and the level of the lesion. Set-up of these patients is similar to that previously described for SSEP and MEP (unless contraindicated). MEPs are not done in cases with cranial abnormalities (large bone flap or metal plate in the skull).

Brainstem Monitoring involves SSEP, MEPs, brainstem auditory evoked responses and free-run EMG from any or all of the following muscles: frontalis, masseter, obicularis oculi, orbicularis oris, masseter, crycothyroid, tongue or trapezius. Triggered EMG is done using either a Kartush bipolar or a Prass monopolar stimulator when requested by the surgeon.

Lipoma monitoring involves MEPs and free-run EMG recorded bilaterally from the anal sphincter, medial hamstring, tibialis anterior and gastrocnemius muscles. Tonic discharges are noted and communicated to the surgeon. Also triggered EMG is done using the bipolar or monopolar stimulator as requested by the surgeon to identify nerve tissue.

For cases with cervical instability median nerve SSEPs are added and occasionally an extra upper limb muscle (median). Baseline traces are obtained on the stretcher with the patient supine and again after positioning on the bed when prone.

Epilepsy Surgery

An electroencephalographer attends every electrocorticography. We record from Ad-Tech single use grid and strip electrodes (most commonly 4x5 and 2x5's in combination). Technologists insert corkscrew recording electrodes for a system reference, ground and reference electrode. Duplicate electrodes are also inserted, in case the original electrode becomes dislodged or faulty during recording.

Once the grid electrodes are placed by the neurosurgeon the technologist plugs them into the 128 channel headbox and the recording is started on a referential montage to a reference on the opposite hemisphere to the surgical site. Photos are taken with an iPad of the patient's position and of the exposed brain prior to and after grid placement, and following the resection. The photo of the patient's position is helpful for orientation of subsequent pictures with grid positions. The photos of the grid on a screen are helpful for discussion in the OR of the contacts involved in epileptic activity. Post resection recordings are occasionally done.

In some cases the electrodes are implanted and the patient returns to the ward for a week of recording. During this time seizures are localized, the sensory strip is identified with SEPs and the motor strip localised with motor stimulation. The patient returns to the operating room for resection of the predetermined area.

Operational Details

All testing is done by technologists who are qualified in multiple modalities. All of our technologists are certified in three or four modalities (EEG, EP, EMG, and IOM), allowing us to provide all diagnostic neurophysiology services in one department. This is advantageous in that all procedures require similar technical expertise and equipment, and gives maximum flexibility in providing services. Technologists also have greater job satisfaction because of the variety and stimulation of their work.

We offer services from 0630-1630 hours on week days and 24/7 on-call coverage for evenings and weekends. We accept routine referrals from all medical practitioners. Requests for specialised diagnostic procedures (epilepsy unit monitoring, ambulatory monitoring, continuous EEG and intraoperative monitoring) require referral from designated neurologists or surgeons.

Technologists and clerical staff work an extended 10-hour day, four days per week. The long hours of service allow staff to be available to match the hours of surgical cases and reduce the frequency of overtime work and call-backs. Call-backs after hours are performed for Neurology or Neurosurgical referrals including infantile spasms, status epilepticus or decreased level of consciousness of unknown etiology.

On a daily basis, we seek ways to enhance the care we provide while continuing to operate efficiently. Technologists take a full history for each patient, and interpretation of EEGs by physicians is first performed without knowledge of the patient's history. The history is taken into account for the clinical impression part of the report only. Such information is saved digitally in our database, and provides useful information for the time of the study, especially for retrospective research studies.

A classification system for EEG findings was developed in-house and has been used for the past 25 years. There is a regular formal review of reports for data accuracy, and weekly departmental meeting are held to identify problems and issues and to discuss solutions. There have also been formal reviews of processes to improve efficiency and departmental retreats to evaluate and change aspects of workflow and function. In combination, the EEG classification and history information available for each EEG test allows useful research like follow-up studies. Currently some 80,000 patients over 25 years are available for database inquiry, thus potentially providing large cohorts for studies like EEG findings (e.g. autism, with over 2,000 patients).

Design and Layout

After 25 years we outgrew our original space at BC Children's Hospital and moved into a larger area. Renovation of this area was made possible with private funds, and we had the privilege of designing our new department from scratch to match our needs.

The department floor plan (see Figure 1. 4,500 sq ft floor space) was designed with all patient testing rooms in one area, separated from the staff and administration area (which includes a large interpretation room, a clerical work area with report storage, offices and conference space). Wide hallways were designed to accommodate patients on stretchers and to provide access for emergency response equipment if required.

There are 9 testing rooms (see Figure 1: 170 sq ft each). Two larger rooms were added (250 sq ft.), one for evoked potential testing (1B28) and the other for EMG and ambulatory/ intensive monitoring set-up (1B20). Each testing room (1B18-28) has a sink just inside the door, millwork cupboards, desk and storage along the back of the room and a stretcher on wheels in the middle of the room. Electrical outlets for the equipment are built into the cupboard millwork at a height of 3 feet off the ground to reduce the number of cables on the floor. A remote control video camera is mounted from the ceiling at the foot of the bed. An electrical outlet is situated at ceiling level for the camera and for an adjacent TV monitor which is mounted high up beside the camera so that when patients are watching a movie, they are looking into the camera. Cables connecting the camera to the EEG machine run through the wall and millwork. Each room has a DVD/video player, patient alarm button, medical suction/air/O2 equipment, telephone, and wall-mounted containers for gloves and sharps.

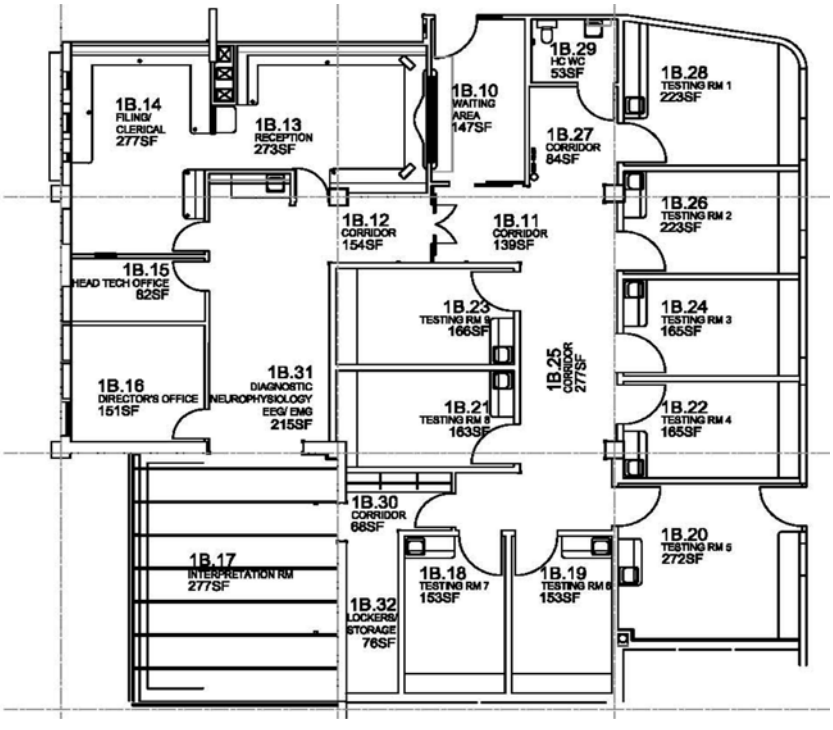

Figure 1. Neurophysiology Floor Plan

In the non-patient area, there is an area for staff lockers, a kitchen sink and fridge. Our interpretation room has three reader stations with network connections to the acquisition systems in the department, the intensive monitoring rooms on the ward (3rd floor) and cEEG recordings in the neonatal or pediatric intensive care units. Another reader station in the conference room is connected to a 70" panel TV for a display of patient data during case presentations. Shelves along one side of this room house our department library, and there are four additional computer terminal workstations with internet access.

Clinical Management System

Our department is highly computerized. We use a customised Clinical Management System (CMS) that manages our technologist resources, appointment bookings, patient demographics, clinical history and EEG report generation. This system allows us to create templates to designate time and resources for various procedures and responsibilities. The technologists' work schedules are entered and based on the number of technologists working each day, appointments slots are opened. Slots are first designated for in-patients, intraoperative work, epilepsy work-up procedures, and weekly rounds or meetings.

Remaining resources are opened for out-patient EEGs, ambulatory monitoring, EPs and EMGs. We routinely have a wait time of >16 weeks for non-urgent outpatient appointments. To accommodate more urgent requests, 3-5 emergency slots are reserved per week. These may be used within a 2-week window for Neurologist referrals. We also have an effective waitlist function to move patients forward when slots become available through cancellations or increased resources. Above and beyond this appointment design, we often receive requests for extra appointments on short notice or for future dates to coordinate with other appointments for out-of-town patients. When possible, we accommodate such requests by using a slot reserved for in-patient testing. We label these "EX" slots so all such requests can be tracked.

CMS is also a very powerful tool for tabulating workload and productivity statistics. Any slot not used for a patient activity (eg. no-show or no inpatient) is coded with the non-patient activity done during that time. This allows tabulation of research and development time spent in different areas.

Archiving and Data Management

Approximately four months of EEG data resides on our network and acquisition stations. Earlier data is archived onto offline external hard drives. Currently such mass storage units are external USB. Each drive has 2 TB of capacity with two mirrored drives in a RAID storage array with redundancy. They connect by USB and can be archived to and retrieved from easily. The files are saved in folders by the month and year. One very large master database on the network contains a list of the date, patient name and archive disk number for all patient recordings.

The proper handling of patient data is an important aspect of management in any department. In our protocol, all EEG/video data is saved locally to the hard drive of the acquisition system and is then copied to a network at the end of the study. The recordings are accessed from the network for interpretation and are then archived offline. An in-house computer program has been used to assist in the data validation. It checks to ensure that all data files have been properly transferred from the collection systems to the network drive. Any files not transferred, missing, corrupted or null-length are identified. In order to keep patient information consistent across different databases, all demographic information is verified with our in-house patient scheduling program. Any mismatches in patient name spelling, birth date or hospital numbers are identified and manually corrected. The data is then archived onto the USB drive and the program is run again to check that the files have been archived 100% properly. Once all EEG/video data is verified, it is deleted from the network drive and the collectors. The same validation procedure is applied to the IM and cEEG data, which is not copied to the network on account of their huge volume, but are archived directly from the collectors.

Education

We foster an environment of continual learning. There are 1-3 Epilepsy Fellows in the department on an ongoing basis, along with a Neurology Resident or Fellow, student EEG technologists and technologists who are upgrading their skills and learning new modalities. Learning occurs via weekly video-EEG rounds, Epilepsy Surgery rounds and reading sessions attended by encephalographers, Fellows, Residents and technologists.

Technologists are often certified in EEG first and then go on to study evoked potentials, EMG and intraoperative monitoring. We have modular teaching presentations for many topics in these areas which are used to prepare for certification exams. They are encouraged to add to their professional registrations.

Student EEG technologists from the British Columbia Institute of Technologist (BCIT) have spent time in our department on practicum placements since 2007. Prior to this this, we offered an in-house EEG Technologist training program which had its inception in 1984. We have extensive outlines, goals, objectives and modular teaching presentations for EEG topics.

Our department has a website with information to educate families about the tests we perform, including preparation instructions and helpful hints. There is also information for referring doctors about the usefulness of and indications for ordering various procedures.

Research

Our patient database is a very powerful tool for research projects. It contains demographics, clinical histories and EEG results for all patients seen in the past 30 years. Clinical information including birth history, development, seizures, neurological

examination, diagnosis, and radiology results are recorded. The EEG report is summarized via a coded classification system. Retrieval of this information is easy and is available for studies and research papers.

Areas of interest over the years have been Benign Rolandic Epilepsy, centro-temporal discharges and the prognostic value of neonatal EEG. Our database has enabled comparison studies involving hundreds of patients with Rolandic dipole vs. non-dipole discharges and neonatal EEGs with normal vs. abnormal background.

Conclusion

The ability to carry out 26 electrophysiological procedures within one department is quite a feat, and one that we are most proud of. It requires dedicated, compassionate and skilled staffs who are qualified and knowledgeable in all modalities. Each member is motivated to work hard to obtain high quality results that will assist physicians in offering the best care for each patient.

The essential factors necessary to run a smooth Neurophysiology program are retaining qualified staff (in an era of a world-wide shortage), having time and motivation to keep skills current, and keeping up-to-date with changes in technology. The common challenges that arise are long wait times for non-urgent tests, competition for allocation of resources to programs with conflicting demands, and difficulty getting support for continuing education. We will continue to work hard together with other departments to meet the needs of our patients and their families, as well as to keep up with the advances in Neurophysiology.

Neurodevelopmental Outcomes after Preterm Birth

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Introduction

Advances in perinatal medicine and neonatal intensive care have revolutionised outcomes for neonates in the past thirty years[1-7]. The widespread use of evidence-based treatments, such as antenatal corticosteroids and surfactant therapy, and other improvements in delivery room resuscitation and nutritional care have resulted in dramatically improved survival, especially among infants born at the threshold of viability[1, 3, 4, 6, 7]. There have been concerns about whether these new survivors face an increased risk of neurodevelopmental disabilities, including impaired cognitive skills, motor deficits, sensory impairment, and behavioural & psychological problems[3, 5, 8, 9]. There is increasing evidence of sustained adverse outcomes into school age and adolescence, not only for extremely-low-birth-weight (ELBW, ≤1000 grams) infants but also for infants born at late preterm[5, 8, 10]. A substantial proportion of "apparently neurologically normal" preterm infants exhibits minor and moderate neuromotor dysfunctions that are not generally detected before school age[11]. The overall adverse short- and long-term health outcomes of these babies can impose a considerable burden on finite healthcare resources[12].

Neurodevelopmental Outcomes After Preterm Birth – An Overview

Preterm birth is an emerging public health problem worldwide. Its prevalence in Canada increased from 6.3% of live births in 1981–1983 to 7.7% in 2010[13, 14]; this is lower than the prevalence in the United States, where preterm birth affected 1 of every 9 infants[15]. The factors cited in connection with this increased prevalence include increased rates of obstetric intervention (i.e. medically indicated labour induction and caesarean delivery) and increases in maternal age and multiple births[13]. Preterm birth is the leading cause of long-term neurological disabilities in children in the United States[15], and it accounts for 45% of children with cerebral palsy[16].

Although survival rates for preterm infants have increased in recent decades, motor disabilities (including cerebral palsy) persist, and impairments in cognitive, language, social, and executive functions have not decreased[17]. The preterm brain develops rapidly during the third trimester, when the volume of the whole brain doubles and the volume of grey matter increases approximately fourfold[18]. This rapidly growing immature brain is exposed to an extremely complicated and stressful environment outside of the womb[19]. Preterm

infants are at high risk for brain injury, which is considered to be a complex amalgam of primary destructive disease and secondary maturational and trophic disturbances[20]. Their clinical manifestations on follow-up can include cerebral palsy, developmental coordination disorder, visual impairment, hearing impairment, cognitive impairment and neurobehavioural disorders[21].

Among the most immature survivors (ELBW or extremely low gestational age neonates, ELGAN), a high prevalence of neurodevelopmental disability has been consistently reported in different cohorts. In the EPICure study from the United Kingdom & Ireland, 308 surviving patients born in 1995 who were less than 25 weeks of gestation were assessed; 30% had developmental delay, defined as scores less than 2 standard deviations (S.D.) below the mean on the Bayley Mental Developmental Index (MDI) and Bayley Psychomotor Developmental Index (PDI), at 30 months of age[22]. Ten percent of the assessed children had severe neuromotor disability, 7% were blind or perceived light only, and 3% had hearing loss that was uncorrectable or required aids[22].

In a large scale study from the United States National Institute of Child Health and Human Development (NICHD) that included 5,250 ELBW infants who were born between 1998 and 2001, <1% of live-born infants weighin ≤500 g survived without impairment (defined as a Bayley score ≥85, a normal neurologic examination, and normal vision, hearing, and walking) at 18 months; this value increased to almost 24% for infants weighing 901 to 1,000 g at birth[23]. Female gender, singleton birth, higher birth weight, an absence of neonatal morbidities, private health insurance, and white race increase the likelihood of unimpaired status in this cohort[23].

A more recent population-based prospective cohort of consecutive extremely preterm infants born before 27 weeks of gestation was conducted between 2004 and 2007 in Sweden, where universal access to antenatal health care and general health insurance is available to all citizens. In that study, the children in the preterm group were compared with control participants in terms of cognition, language, and motor functions at a median age of 30.5 months using the Bayley-III scales[24]. Overall, 16% had moderate disability (defined as scores between -2 and -3 S.D. from the mean of any of the Bayley-III scales, able to walk with aids, hearing impairment corrected with a hearing aid, and registered at low vision centres without blindness), and 11% had severe disability (Bayley-III composite cognitive, language or motor score < mean – 3 S.D., unable to walk even with an aid, or bilateral blindness or deafness).

In the EPIPAGE study, which included follow-up of 2,901 livebirths between 22 and 32 completed weeks of gestation from nine regions in France in the post-surfactant era, the children underwent a medical examination and a cognitive assessment with the Kaufman assessment battery for children (K-ABC), and their scores on the mental processing composite (MPC) scale were recorded at 5 years of age[25]. Cerebral palsy was diagnosed

in 159 (9%) of the children born very preterm; 503 (32%) had an MPC score less than 85, and 182 (12%) had an MPC score less than 70[25]. The likelihood of having cognitive and neuromotor impairments at 5 years of age increased with decreasing gestational age, and many of the children born very preterm required a high level of specialised care.

A meta-analysis that included 9,653 children with a mean birth weight of 1,060 g and a mean gestational age of 28.2 weeks in 41 studies demonstrated clear evidence of substantial motor impairments in very preterm or very-low-birth-weight (VLBW) children from infancy to 15 years of age[26].

Very preterm and/or VLBW children also have moderate-to-severe deficits in academic achievement, attention problems, and internalizing behavioural problems and poor executive functions, and continue to lag behind their term-born peers during their transition to young adulthood[27]. In the follow-up of the EPICURE cohort, 30% attended special schools, and approximately 60% of those attending mainstream schools had special education needs at 11 years of age[28]. Upon follow-up, these extremely preterm children were more than three times more likely than their classmates to have a psychiatric disorder, including attention-deficit/hyperactivity disorder (ADHD), emotional disorders, and autism spectrum disorders[29].

Fine and gross motor coordination deficits are not uncommon among survivors born preterm. Developmental coordination disorder, defined as impaired motor performance sufficient to produce functional impairment that cannot be otherwise explained by the child's age, cognitive ability, or neurologic or psychiatric diagnosis, is found in 31% to 34% of VLBW and 50% of ELBW infants[30].

There are many challenges in interpreting neurodevelopmental outcome studies because of their heterogeneous nature, which includes differences in survival and neurodevelopmental impairment rates by geographic region and neonatal network related to multiple factors, including population characteristics, perinatal/neonatal management, follow-up protocols, assessments, and even definitions[30]. The Bayley Scales of Infant Development is not without limitations, despite being the most widely used measure to assess neurodevelopment in very preterm and VLBW infants in the first three years of life. In 2006, the newly revised version (Bayley-III) was introduced in an attempt to isolate cognitive skills from language skills[31]. However, the Bayley-III cognitive scores were significantly higher than Bayley II MDI scores for term and preterm toddlers, both combined and separately, which implied a need to exercise caution when using test scores from different versions of a test as the primary outcome measure in research studies[31]. Moreover, a recent meta-analysis showed a limited predictive value of Bayley Scales of Infant Development scores for the later development of preterm infants, with the MDI explaining 37% of later cognitive functioning and the Bayley motor scale explaining 12% of later motor functioning[32]. Further research on developmental assessment tools with sufficient sensitivity and specificity to predict later cognitive and motor development and guide future interventions is needed[32].

Prediction Of Neurodevelopmental Outcomes – Imaging Techniques

Cranial ultrasound (CUS) has been routinely used as the primary imaging tool for evaluating brain injury in premature neonates since its introduction in 1980s[27, 33]. It is non-invasive, inexpensive and portable, and it allows the accurate detection of intraventricular haemorrhage (IVH), cystic periventricular white matter injury and hydrocephalus[34]. In the 2013 Canadian Neonatal Network report, 9.7% of infants with a GA<33 weeks exhibited neuroimaging abnormalities, with 15%, 7% and 3% presenting ventricular enlargement, intraparenchymal lesions and periventricular leukomalacia (PVL), respectively[35]. Among VLBW infants, 48% developed varying degrees of IVH, ranging from germinal matrix haemorrhage to grade 4[35]. In the past, small haemorrhages (grade 1 or 2 IVH) were believed to not increase the risk of disability significantly beyond the background risk associated with gestational age and birth weight[27]. However, current evidence suggests that even a small IVH increases the risk of disability in terms of cerebral palsy or neurodevelopmental impairment, even in infants with only grade 1-2 IVH[36, 37].

CUS has a limited ability to accurately diagnose non-cystic white matter injury and other brain parenchymal injuries[38]. For example, diffuse PVL has been regarded as an important cause of cognitive deficits and chronic neurodevelopmental impairment, but it is difficult to recognise with CUS[39, 40]. Even serial CUS only has a sensitivity of 26% for identifying white matter abnormalities in premature infants[41]. Advanced neuroimaging methods, such as magnetic resonance imaging (MRI), magnetic resonance spectroscopy, and diffusion-weighted MRI, have identified patterns of damage after insult to the immature brain[42]. The brain injury patterns identified and the regional changes observed on MRI are predictive of particular neurodevelopmental syndromes later in childhood[40, 43].

White matter abnormalities, including parenchymal lesions and punctate hyperintensities, are typically readily identifiable with conventional imaging sequences, including T1- and T2-weighted images, and their developmental effects have been studied extensively[34]. Parenchymal lesions on term-equivalent MRI, defined as parenchymal haemorrhage, PVL, infarction, and white matter loss, have been reported to have 100% sensitivity and 79% specificity for predicting cerebral palsy at 18 months of age[44]. In another study conducted by Kanagawa Children's Medical Centre in Japan, all 37 patients with cerebral palsy who were evaluated at 3 to 5 years of age had periventricular lesions with T1 hyperintensity or cysts in the corona radiata above the posterior limb of the internal capsule on coronal sections, and there was a tendency for the presence of widespread lesions in this region to be correlated with the severity of the motor handicap[45]. In a cohort of infants born at <34 weeks gestational age, an abnormal neurodevelopmental outcome at 18 months adjusted age was associated with increasing severity of white matter injury, ventriculomegaly, and IVH on MRI and moderate/severe abnormalities on the first and second MRI studies, performed at the median ages of 32 and 37 weeks, respectively[46]. Using the Griffiths Mental Development Scales for assessment, Dyet reported that adverse outcomes at 18 months' corrected age were associated with major destructive lesions and diffuse excessive high signal intensity within the white matter[47]. In

contrast, cerebellar haemorrhages observed on MRI studies did not demonstrate a consistent relationship with adverse neurodevelopmental outcomes across different studies[48, 49].

Standardised scoring systems for the results of conventional imaging sequences have been developed to evaluate and quantify regional and/or global injury and provide a comprehensive assessment of cerebral alterations that correlates with neurological outcomes[34]. Significant white matter abnormality quantified by the standard scoring systems not only correlates with adverse early neurodevelopment outcomes[40, 46] but is related to motor and cognitive impairment in childhood[50, 51]. Newer imaging techniques such as volumetry, surface-based morphometry, diffusion tensor imaging and magnetic resonance spectroscopic imaging provide novel insights into aberrant cerebral development in high-risk preterm infants[34, 52]. Recent advances in magnetoencephalography analysis presents an excellent opportunity to map neural processes before and after birth in infants at high risk for disabilities, which may carry prognostic values in their neurodevelopmental outcomes[53-55].

The Neonatal Follow-Up Programme, Vancouver

The Neonatal Follow-Up Program at the British Columbia Children's & Women's Hospital was started in 1983 by Dr. Michael Whitfield to collect audit information about the survival of different categories of high risk infants in the province. The goals of the programme are to evaluate the short and long-term results of perinatal/neonatal intensive care in the Provincial Tertiary Perinatal Unit via sequential clinical and neuro-developmental assessment of surviving infants during infancy and early childhood; to identify impairments early in this high-risk population to promote early intervention that can minimise the severity of perinatally acquired handicaps; to provide educational experience with developmental assessment and the long-term effects of perinatal/neonatal intensive care for learners in perinatal/neonatal care training programmes; and to promote and conduct research to advance knowledge of the long-term effects of select aspects of perinatal and neonatal management.

The programme is multidisciplinary and provides comprehensive, age-appropriate, sequential neurodevelopmental assessments at 4 months, 8 months, 18 months, 3 years, and 4 ½ years corrected age [Table 1]. The clinic staff includes nurses, psychologists, physiotherapists, occupational therapists, audiologists, speech-language pathologists, and paediatricians/neonatologists, in addition to a data manager and office staff.

Patients are recruited for follow-up based on perinatal/neonatal risk factors. The programme focuses on children at highest risk of disability or for whom there is little knowledge about potential outcomes. Current recruitment criteria include birth weight ≤800 g; gestational age ≤25 weeks (for gestational ages 26-28 weeks, one assessment is provided at 18 months [Canadian Neonatal Follow-Up Network]); infants with intraparenchymal/ intracranial haemorrhage, cystic PVL, severe retinopathy of prematurity (grade 4 or treated with laser/Avastin), discharge on home oxygen, congenital diaphragmatic hernia and patients participating in funded studies.

The programme has conducted and been involved in a large number of studies, which vary from funded large international randomised controlled trials, national studies, western Canadian studies and institutional studies to unfunded, often trainee initiated, research studies and comprise obstetrical, neonatal and neonatal neurological studies. The largest ongoing multicentre trial is the 11-year-old assessment of children in the Caffeine for Apnoea of Prematurity. The programme contributes to the Canadian Neonatal Follow-up Network study and the western Canadian Complex Pediatric Therapies registry. Working in close collaboration with our paediatric neurology group, MRI neuroimaging studies have been used to recruit children with congenital heart disease, neonatal encephalopathy and approximately 200 preterm children who have undergone sequential longitudinal assessments in the programme. In 2012-2013, programme staff were investigators on 9 large funded research grants (including 6 on which a programme staff member was the principal investigator) and have published 31 manuscripts and 41 abstracts related to follow-up.

In summary, despite advances in perinatal & neonatal care, surviving preterm infants are at risk for adverse neurodevelopmental outcome. Newer imaging techniques demonstrate an improved ability to predict the development of adverse neurodevelopmental outcomes. A well-organised follow-up programme is essential to identify impairments early in this high-risk population to promote early intervention and to conduct research to advance knowledge of the long-term effects of select aspects of perinatal & neonatal management.

Table 1. Neonatal Follow-up Programme

Discipline	4 Months	8 Months	18 Months	3 Years	4.5 Years
Target Areas	Feeding, Growth and Parental well being	Detect severe impairment. Early intervention	Detect major impairments	Detect minor impairments Community resources	School entry assessment
Pediatrician	✔	✔	✔	✔	✔
Nursing	✔	✔	✔	✔	✔
Physiotherapy		Bayley-III MAI	Bayley-III PDMS-II	Bayley-III PDMS-II M-ABC Sensory profile*	
Occupational Therapy	Feeding Bayley-III MAI	Bayley-III MAI			PDMS-II M-ABC Sensory Profile*
Speech/ Language				PLS-5	PLS-5
Audiology		Behavioral audiometry		Ear specific audiometry	
Psychology					WPPSI-IV VMI WIAT-III BRIEF-P* CBCL*

Tools

1. Bayley-III-Bayley Scales of Infant and Toddler Development 3rd ed
2. MAI-Movement Assessment Index
3. PDMS-II-Peabody Developmental Motor Scales – 2nd ed
4. M-ABC-Movement Assessment Battery for Children – 2nd ed
5. WPPSI-IV-Wechsler Preschool and Primary Scale of Intelligence – 4th ed
6. BRIEF-P-Behavior Rating Index of Executive Function – Preschool ed
7. Sensory Profile-Sensory Profile-Child Sensory Profile 2nd ed
8. PLS-4-Preschool Language Scales 4th ed.
9. VMI-Beery-Buktenica Developmental Test of Visual Motor Integration
10. CBCL-Child Behavior Checklist
11. WIAT-III-Wechsler Individual Achievement Test – 3rd edition

*Questionnaires

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Psychology Services on the Neurology Multi-disciplinary Team

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Case example:

Julie is a 13-year-old girl with a two-month history of seizure-like episodes. She came to British Columbia (BC) Children's Hospital for a full medical work-up. All tests were negative and the results were normal (video EEG monitoring indicated no abnormal brain activity during the seizure-like episodes).

Psychology was consulted. Psychological assessment indicated that there had been significant stress in the family and for the girl over the past year.
Stressors:

- *Mom was in a car accident earlier in the year.*
- *Julie had experienced significant on-line bullying by peers at school.*
- *Julie was very anxious about starting high school next year.*

Initially, Julie and her parents under-reported the significance of these stressful events. It took a trusting relationship, gradual engagement in the social-emotional assessment, and education to help Julie and parents identify the importance of these stressful experiences and to become aware of their impact on Julie's well-being.

Treatment involved:

- *Close collaboration between the neurologist and psychologist in the process of differential diagnosis*
- *Education for Julie and her parents to help them understand that Julie's seizure-like episodes were real, but were not related to epilepsy or another brain-based illness or injury (e.g. a tumour)*
- *Education for Julie and her parents about the mind-body connection*
- *Therapy for Julie to help her be more aware of her emotions and stress and to develop healthy ways of expressing negative emotions in therapy and at home*
- *Liaising with school to help resolve the on-line bullying*
- *Setting up a smooth transition to high school*

Julie recovered over the next 4 months.

The Role of Clinical Psychology

In the multi-disciplinary Neurology service at BC Children's Hospital, psychologists are active members of the team, collaborating with the other health professionals to help with differential diagnosis (as in the case example above), improving coping and compliance with a medical condition and its treatment, and providing support around the social-emotional and behavioural sequelae of neurological conditions. The treatment offered by the psychologist includes education for the child and the family on mind-body connection, common challenges children with neurological conditions may face in every day life, and how to utilise effective coping strategies to manage the stress associated with the illness. The identification of stressors plays an important role in promoting stress management. When the child and parents begin to appreciate the role of stress in symptom management, they are more likely to accept and utilise stress management tools such as relaxation training.

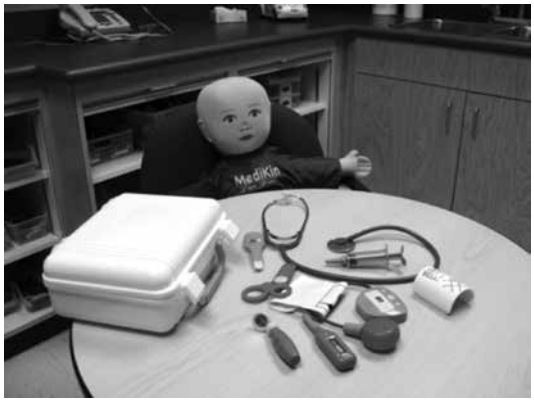
For more information on stress management, please go to:
http://www.bcepilepsy.com/files/Information_Sheets/Stress_Management.pdf

Parents play a key role in promoting the social-emotional functioning of their children with neurological problems. The psychologist:

- Works with parents to effectively cope with the diagnosis
- Provides parent education on managing symptoms and promoting normal development (e.g. independence, social relationships, learning)
- Educates parents on parenting their children with neurological conditions (e.g. epilepsy) during developmental transitions (e.g. being not too restrictive when the child grows from childhood to adolescence)

The psychologist also provides support to connect the child to community resources. In addition to connecting the family with counselling in the community, a psychologist may also liaise with school or recreational/social professionals to support the child's smooth transition back to community-based activities. The goal is to ensure the optimal functioning of the child in the community. Parent and child education on symptom management and planning for potential challenges will empower the child and family to effectively cope and live with the neurological condition. It feels less frightening for the child and family when they know what they can do to cope when pain is high or when the child is having symptoms.

Different psychological approaches are used to assess and treat children with medical issues. A social-emotional assessment is typically used to identify stressors and explore coping styles. Play therapy or cognitive behavioural therapy (CBT) is used to treat the child's social-emotional challenges and promote coping. Stressors in the family system are addressed with parents or with the whole family. If a child needs hospitalisation, the psychologist works with Child Life specialists to promote the child's coping with procedures and surgeries during the in-patient stay. The psychologist also works with the school to support the child's functioning and learning. In the unfortunate situation of a child death, bereavement services are provided.



The Role of Neuropsychology

A neuropsychologist's role on the Neurology multi-disciplinary service is to provide assessment and consultation regarding a child's cognitive development and functioning in the context of the neurological condition. The neuropsychologist has specialised expertise in identifying the relationship of brain-based illnesses or injuries to problems in thinking, learning, emotional, and behavioural functioning in everyday life. Neuropsychological assessments inform the medical team about the impact of the health condition on the child's functioning and assist parents, teachers, and other community-based professionals in supporting the child's optimal development and function.

At BC Children's Hospital, a child may receive neuropsychology assessment when there is concern about how a neurological condition and/or its treatment is affecting a child's cognitive development or function. All children receiving neurosurgical treatment for medically-refractory seizures have a neuropsychological assessment prior to surgery and serially for 5-10 years after surgery.

Common reasons for referral for a neuropsychological assessment are:

- To provide diagnostic assistance or clarification (i.e. is the child's neuropsychological profile of strengths and weaknesses consistent with what would be expected of the condition?)
- To establish a baseline before treatment (e.g. before neurosurgery, before the start of a new drug)
- To track progression of illness and/or response to treatment (i.e. are there improvements or declines in neuropsychological functioning over time?)
- To describe/monitor cognitive functioning in the context of the medical condition (i.e. does the neuropsychological profile suggest localised or lateralised dysfunction? Does the child have a specific cognitive deficit or learning disability as a result of the medical condition?)

The neuropsychologist provides consultations to the medical team, parents, and teachers/schools about the impact of the health condition on the child's development, thinking/cognition, social-emotional functioning, and functioning in school with specific

emphasis on how illnesses and injuries of the brain can affect daily functioning. Often the neuropsychologist collaborates with school to provide specific information about the learning profile of children with specific neurological conditions and to assist in development of special education services and intervention to support the child in the classroom.

The following photographs show some of the tools used.

The EEG Technologist training programme

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British Columbia Children's Hospital's (BCCH) first opened its doors in 1982 in the current site. As the only tertiary care centre for paediatrics in the province, the hospital sees children with a wide variety of presenting complaints. At its inception, the Department of Diagnostic Neurophysiology had 5 full-time technologists performing EEGs (electroencephalography), one of whom also performed EMGs (electromyograhy). Currently the department has 11 full-time and 1 part-time multidisciplinary technologists who are certified in EEG, EMG, EPS(evoked potentials) and intraoperative neuromonitoring. Six of those technologists are graduates of the Children's Hospital training programme. Each member of our department is dedicated and motivated to obtain high quality results that will assist physicians in offering the best care for the patient.

History Of Recruitment For Our Eeg Training Programme

In 1984 the department decided to establish its own 2-year in-house EEG training programme with the intention of training technologists to take on full-time positions in the department. In this way, students could be hand-picked and trained to meet the specific demands of the department.

Students were initially recruited through the hospital's volunteer programme newsletter. Applicants included high-school students who were volunteering to get experience in a paediatric medical environment prior to graduating and going into the medical field, and older applicants who had an interest in paediatrics and were looking for a career change. Requirements for application were simple: graduation from high school with a C+/B average. After the first applicants successfully completed the programme and were hired, the recruitment process was expanded to include local high schools. Flyers advertising the programme were circulated around the local high schools at graduation time and when possible, a technologist would present an overview of the programme on career day.

At this time we also expanded our training to include students that had graduated from the 2 year full-time electroneurophysiology diploma programme at the British Columbia Institute of Technology (BCIT). Graduates of this programme require more patient/clinical experience in order to sit their Canadian Board of Registered EEG Technologists (CBRET) final examination. For those students we offered a modified one-year programme covering the more advanced syllabus that students in the BCCH programme would receive in the second year of their training.

In more recent years our recruitment and requirements for applicants has changed significantly. Applicants must have a B or greater graduating average from high school with a strong emphasis on science courses. Applicants must also have recently graduated from a degree or diploma programme with a 75% or greater average. Preference was given to graduates in the field of sciences who had work experience (not necessarily related to the medical field or paediatrics). Recruitment is solely based in the local universities. The disadvantages of this recruitment philosophy are that applicants are older and in most cases living independently from their family. Many have incurred student loans and other debts. Unlike applicants who were straight out of high school and still living with family, these applicants often cannot financially afford to be in a two-year training programme with only a minimum wage pay scale. In addition, many have a long-term goal of entering medical school and see our programme as a "stepping stone" towards that goal. These applicants do not fit with our philosophy of training students who wish to be employed in our department and who see diagnostic neurophysiology as a long-term professional commitment.

We currently also offer our department as a practicum site for BCIT students who are looking for a department where they can obtain the minimum number of paediatric patients that are required in order to sit the CBRET final examination. These students are here strictly to perform EEGs under the guidance of a qualified technologist. If there are not enough suitable patients in a day, and time permits, they may have the chance to review their records with a technologist. Employment in our department is not contingent with this service.

The Programme

The two year programme covers practical training in the set-up and running of an EEG, the interpretation of EEG data as well as in-depth study of electronics, neuroanatomy and clinical disease processes including, but not limited to, neurological and epileptic disorders. All practical training is done under the direct supervision of a qualified technologist. All tests and lectures [see Table 1] are prepared and given by a qualified technologist.

Table 1

Lectures 1 – 3 Months	Lectures 4 – 6 Months
10-20 measuring system	electronics
running a record	digital EEG theory
patient safety	electrodes
normal waking and sleep	full calibration
EEG convention and montages	function of lobes
artifacts	specific seizure types and epileptiform waveforms
activation procedures	classification of epilepsy and basic epileptic syndromes
normal variants	
abnormal non-epileptiform waveforms	
basic seizure types & non-specific epileptiform waveforms	
EEG classification system	

Lectures 7 - 9 Months	Lectures 10 - 12 Months
pharmacology	electrical safety
brainstem and cranial nerves	myoclonus
neurological exam	generators of EEG
patient history	infectious diseases and infection control
basic anatomy of cerebellum, basal ganglia, pituitary gland, internal capsule and diencephalon	anoxia and coma
skull and meninges	ECS
ventricular system, CSF and hydrocephalus	degenerative diseases
sleep staging and disorders	dementias
cerebral circulation	metabolic disorders
vascular diseases / disorders	trauma and head injuries
headaches and migraines	respiratory systems, apnea and breathholding spells
tumours	
Lectures 13-24 Months	
neonatal EEG	
development of the brain and spinal cord	
spinal cord anatomy	
auditory and visual systems	
neuroimaging	
epileptic syndromes and status epilepticus	
chromosomal and congenital anomalies	
psychiatry	
autonomic and limbic systems	
peripheral nerve disorders	

The First Year

The first year of the programme is divided into four 3-month blocks. During each of the block a student is paired with a senior technologist "teacher". At the beginning of each block the student receives a set of practical goals that they need to meet. At the end of each block written test(s) is /are given to assess the student's understanding and competency. Each day of the block is comprised of two EEGs (1.5 hours total time for each) during which practical applications and history taking are the focus. Each EEG is followed by a 1.5 hour session during which EEG interpretation is the main focus. During this time they review the patient's history, determine what EEG findings would be expected in that clinical scenario and what technical factors should be taken into consideration (i.e. activation procedures, additional electrodes, etc.). Lectures and tests are given once a week with tests encompassing the entire syllabus covered up to that time. Towards the end of the first year more multiple choices questions are introduced in preparation for the CBRET written examination.

1-3 months:

The objectives during this time are head measurement and electrode application, basic EEG concepts and the identification and interpretation of basic EEG waveforms. Lectures during this block focus on the International 10-20 system, basic recording strategies/ techniques and patient safety, activation procedures, basic waveform identification (normal and abnormal waking and sleep), identification and troubleshooting of both physiological

and non-physiological artifacts, basic seizure types, basic digital instrumentation, montages and EEG convention.

At the end of this block, the student is expected to be able to measure and apply a set of electrodes using the 10-20 system in one hour or less with fewer than 5 errors (an error is defined as any inter-electrode measurement that is incorrect by more than 5 mm). The student should also be able to correctly identify all the normal waking and sleep waveforms, identify all the sleep states and have a good understanding of the identification and correction of artifacts and technical errors (i.e. incorrect inter-electrode distances).

4-6 months:

The objectives of this block emphasize hands-on learning including more efficient electrode application, running an EEG with the assistance of the teacher and practicing waveform identification while running an EEG. Lecture material contains topics on more advanced electronics, more advanced digital instrumentation, electrode properties, neurophysiology, specific seizure types/classification, epileptic waveforms, classification of epilepsy and basic epileptic syndromes.

At the end of this block the student is expected to measure and apply a full-set of 10-20 electrodes on a co-operative patient in less than one hour with no more than 3 errors. In addition, the student should be able to independently run a normal EEG recording or one with only very minor, non-complicated abnormalities.

7-9 months:

This block focuses on the student's ability to run EEGs on older, co-operative patients independently. At this time the skills necessary to perform EEGs on younger children (including measuring and applying electrodes on patients while they are lying down and moving) are introduced. Lectures during this block have a stronger emphasis on clinical theory. Topics include taking a detailed clinical history, neuroanatomy, basic neurological disorders and their effects on the EEG, the neurological examination, sleep staging and disorders.

At the end of this block the student is expected to be proficient in taking a detailed clinical history and measuring and applying a full set of 10-20 electrodes in less than 45 minutes on a co-operative older patient with no more than 3 errors. They should also be able to independently record the EEG and remove the electrodes. The objective is to accomplish all of this in 90 minutes or less.

10-12 months:

The objectives for the final three months incorporate portable EEG recordings and dealing with more critically ill patients. Lectures cover a wide range of subjects including electrical safety, infection control, complex neurological disorders and their EEG presentation, respiratory systems and abnormalities arising from dysfunction. The guidelines for electrocerebral silence (ECS) recordings and the recognition of artifacts in those situations is covered in detail.

At the end of this block the student is expected to be proficient in taking a history on any type of patient and should be proficient in measuring and applying electrodes in less than 30 minutes with no more than one error. Additionally, they should be able to routinely take a complete history, measure and apply the electrodes and run the recording (including all of the appropriate activation procedures and the appropriate wait for sleep should it not be obtainable) in 90 minutes or less on any patient.

The Second Year

This year focuses on preparation for the CBRET practical examination. CBRET requires that students must have trained for a minimum of 24 months under the direct supervision of a qualified technologist. Candidates are required to perform and document 500 EEGs in patients from a wide age spectrum. In a training programme such as ours, based in a paediatric facility, 400 of those EEGs can be performed on children (age 12 years or less), 100 must be recorded on adults (21 years or older) with 20 of those being greater than 60 years of age.

During the second year the student continues to work under the supervision of a qualified technologist, however they perform EEGs independently. Prior to completing a patient and removing the electrodes the record is reviewed by a senior technologist. If all of the technical requirements have been met and the appropriate states captured the student may then remove electrodes and clean up the patient. The student performs 3 EEGs a day with time set aside at the end of the day to review the records with a technologist. During this review time emphasis is placed on oral examination techniques, correlation of the presenting history with the EEG findings and recognition and definition of both normal and abnormal findings.

Lecture topics during the second year are more advanced and include neonatal EEG, development of the brain and spinal cord and in-depth neuroanatomy. Peripheral nerve disorders, neuroimaging, psychiatry, status epilepticus and epileptic syndromes are also covered. Evoked potential basics are introduced in the last 3 months of training.

At the end of the 2-year programme, the candidate will be independent and confident in the role of neurophysiology technologist.

Conclusion

There are many challenges to run a 2-year intensive training program. The first is to gain financial approval from the hospital to hire a student. Another financial consideration is that the full-time employee capacity is reduced due to one senior technologist being dedicated to the training programme. Although there are difficulties in recruiting a student, once you have found an ideal candidate and they have completed the course you have the benefit of a fully trained, compassionate and knowledgeable neurophysiology technologist, ready to take an active role in the field of neurophysiology.