Esophageal Atresia and Tracheoesophageal Fistula (EA/TEF) is a congenital birth defect that occurs in one in every 2,500 to 3,000 babies. In most cases, this defect occurs randomly but a small percentage of cases are linked to chromosomal abnormalities, or a genetic disorder.

The EA/TEF Clinic of The Montreal Children’s Hospital of the McGill University Health Centre provides EA/TEF patients and their families with multidisciplinary and interdisciplinary expertise to optimize the care and prevent the morbidity associated with complications that may develop during follow-up. All newborns with this malformation are automatically referred to the clinic and followed according to a well-defined protocol. Older children and adolescents who were previously operated for the malformation can be evaluated in the clinic to ensure standardization of follow-up, and preparation for a smooth transition to a specialized adult site in the future.

Signs of EA/TEF to watch out for in a foetus or neonate include:

- A large amount of amniotic fluid (also known as polyhydramnios).
- Once born, the baby has trouble swallowing his/her saliva; bubbly saliva may be seen coming out of the baby’s mouth.
- The newborn may have a rattling respiration and episodes of coughing and choking.
- The baby may appear cyanotic and may have an enlarged abdomen due to air building up in his/her stomach.

For more information:

**EA/TEF Clinic**

Tel.: 514-412-4400, ext. 23435
The Montreal Children's Hospital Nephrology Division, under the directorship of Dr. Martin Bitzan, has added a number of novel extracorporeal treatments (ECT) to its roster of services and there are plans to continue developing its expertise in the field.

Besides conventional hemodialysis, some of the new services available include various forms of continuous renal replacement therapy (CRRT) that can be combined with other procedures, such as extracorporeal membrane oxygenation (ECMO).

In addition, the MCH has recently started ECT for non-kidney disease in the form of plasmapheresis or plasma exchange. For example, the team now offers ECT for several neurological diseases such as myasthenia gravis, an acquired neuromuscular disease leading to muscle weakness and fatiguability, caused by circulating antibodies that block acetylcholine receptors at the post-synaptic neuromuscular junction. “This ECT intervention is an end-of-line therapy for autoimmune diseases, employed when no other treatment works, in conjunction with standard therapies,” says Dr. Michael Zappitelli, Director of the Extracorporeal Blood Therapy and Peritoneal Dialysis Program in the Nephrology Division. “This intensive treatment and others we provide (like CRRT) require highly specialized nursing care and specialized machines and are often provided to very sick children. Our hemodialysis and critical care nurses are developing a niche in these particular treatments. These services are rarely used for children, but thanks to our growing expertise, our patients no longer have to be transferred to an adult hospital for the treatment.”

Looking down the road, the division hopes to continue expanding its ECT program if there is demand. Potential new services include red blood cell exchange for children with sickle cell disease; white blood cell depletion for children with acute leukemia; phototherapy for various cancers; and retrieval of blood stem cells. While the institutional need and feasibility for these novel services is yet to be established, Dr. Zappitelli is confident that the MCH nursing and medical staff have the knowledge and personnel to adapt and provide state-of-the-art pediatric ECT.

For more information:
MCH Nephrology Division
514-412-4400 x 24461

Pauline Espinosa, Assistant Head Nurse
Hemodialysis, and Alex
Diabetes is a complex disease, with more than 47 genes already involved and more to come according to Dr. Constantin Polyzonacos, director of the Division of Pediatric Endocrinology at The Montreal Children's Hospital. Rather than being discouraged, however, Dr. Polyzonacos sees the identification of each new gene as one step closer to a cure.

A case in point is his latest findings, which have revealed the gene responsible for a very rare form of diabetes, affecting children who are born with a congenital absence of the cells that make insulin in the pancreas. Their study indicates that the gene RFX6 plays a major role in the development of these cells. They postulate that this gene is a key player in converting exocrine cells that do not secrete insulin into islet cells that do. This serendipitous discovery has major implications for both Type-1 and Type-2 diabetes, where the ideal treatment is transplantation. Scarcity of donated pancreatic material, low success rates and graft rejection are limiting factors to this procedure. With the discovery of the RFX6 gene, these problems may no longer be an issue since it may be possible to use RFX6 to produce islet cells for transplantation. Procedures such as using gene therapy or tissue culture conversion of cells into islets followed by auto-transplantation hold promise. The next steps for Dr. Polyzonacos and his team will be to validate the ability of this gene to drive islet cell production and to use it as a marker of disease.

Genetic and genomic research can seem overwhelming since there are so many genes involved in complex diseases such as diabetes, but Dr. Polyzonacos latest research clearly demonstrates the importance of tenacity: one gene can tip the balance between treatment and cure. “This is the main reason I’m doing genetic research,” he says. There are more than 170 million diabetics worldwide.

For more information:  
**Endocrinology Division**  
of the Montreal Children’s Hospital, McGill University  
and McGill University Health Centre  
Dr. Constantin Polyzonacos, MD-PhD  
Professor, Department of Pediatrics  
Tel.: 514-412-4400, ext. 22482  
email: constantin.polyzonacos@mcgill.ca
Cancer Research

A common pathway may be key to treatment of pediatric astrocytomas

The world stops turning for parents whose children have been diagnosed with a brain tumour. “The brain controls everything in our body including our intellect. Parents who are given a diagnosis of a brain tumour no longer see their children as having the chance to be ever normal again,” says Dr. Nada Jabado, a researcher at The Montreal Children’s Hospital of the McGill University Health Centre, who is studying the molecular events involved with the development and progression of pediatric astrocytomas, a group of non-neuronal brain tumours.

Pediatric astrocytomas account for 45 percent of all pediatric brain tumours. Children with high-grade astrocytomas have a three-year survival rate that is less than 20 percent.

Dr. Jabado’s research focuses on identifying the signaling pathways of both high- and low-grade astrocytomas using biochemical, gene and genome profiling in primary tumours, cell lines and mouse models. She is also interested in pinpointing common pathways used by these tumours and present in other cancers, as this has shown clinical promise. For example, she and others have shown that RAF and the MAP kinase pathways play pivotal roles in low-grade astrocytomas.

Findings on high-grade tumours have shown that they are more homogenous than previously thought, and based on the identified altered events/pathways in these subsets, there may be treatment options already available. Determining the effectiveness of and extending available therapies is particularly relevant for children since current treatments involve chemotherapeutics and radiation therapy, both of which have side effects including potential damage to the developing brain. Children who are too young to receive radiation therapy or have already failed some chemotherapeutics need additional therapeutic options. Characterizing the key pathways involved in these tumours has also led to the discovery of new drug targets, which now need to be validated.

Another important benefit of this research will be to identify patient subgroups that will respond to particular therapies. This personalized approach will reduce the use of nonfunctional treatments.

For more information:
Hematology-Oncology Division of the Montreal Children’s Hospital
Dr. Nada Jabado, MD-PhD, Assistant Professor, Department of Pediatrics
Tel.: 514-412-4445
email: nada.jabado@mcgill.ca

Perinatal Grief Program

In 2000, the Vaudreuil-Soulanges CSSS implemented a comprehensive prevention program involving personalized, individual follow-up and a support group (Paroles aux anges) for bereaved parents who have lost a pregnancy or a newborn child. In October 2007, the program was awarded a Prix d’excellence du réseau de la santé et des services sociaux [health and social services excellence award] in the category Personnalisation des soins et des services [personalized care and services].

The program’s main objectives include breaking through grieving parents’ isolation by encouraging them to express their feelings, teaching them about the perinatal grieving process, bringing perspective to their experience, providing specialized documentation, screening for and preventing mental health problems, and screening for and preventing attachment issues with a subsequent baby. The program also provides support to workers involved in these cases.

In June 2006, the Vaudreuil-Soulanges CSSS designed a set of tools for institutions wanting to set up this type of program. These tools help to provide more standardized perinatal bereavement services in healthcare institutions, and are an effective and inexpensive training resource for all healthcare workers who wish to work with bereaved families and raise awareness among colleagues. To expedite the process, a CD ROM containing a number of files is included and can be used to customize the tools to individual institutions and based on available resources. Workers can therefore focus all of their energies on helping clients rather than losing valuable time redesigning existing tools.

To compensate for the lack of documents available to bereaved parents, two brochures, available in both French and English, were produced in June 2006. Clients have expressed their overall satisfaction with our brochures, in terms of both the relevance of their content and their attractive, easy-to-use format. Everyone agrees that these brochures, which contain relevant, reassuring and soothing information, should be given to any grieving parent after the loss of a pregnancy or a newborn child.

For more information on these resources, contact
CSSS de Vaudreuil-Soulanges
Manon Cyr, R.N. specialized in perinatal bereavement
Tel.: 450 455 6171, ext. 359
Email: manon_cyr@rrsss16.gouv.qc.ca
CONTINUING EDUCATION

Pediatric Outreach and Telehealth Programs
Location: MCH and by videoconference

LES TUMEURS CÉRÉBRALES PÉDIATRIQUES
(presentation in French)
Date: September 8, 2010 from 1:30 to 3:00 p.m.
Description: Revue de l’anatomie du cerveau, les types de tumeurs pédiatriques et les traitements: impact sur la famille

FACILITER LA RÉINTÉGRATION SCOLAIRE DE L’ENFANT APRÈS DES TRAITEMENTS CONTRE UN CANCER
(presentation in French)
Date: October 13, 2010 from 1:30 to 3:00 p.m.
Description: Revue des écrits scientifiques sur le sujet de la réintégration scolaire après des traitements contre le cancer. Discussion sur les stratégies infirmières qui aident l’enfant et sa famille à la réintégration scolaire.

Trauma Rounds
Location: MCH amphitheatre and by videoconference

TRAUMA TEAM ACTIVATION AT THE MCH: A 4-YEAR REVIEW
(presentation in English)
Date: September 27, 2010 from 8:00 to 9:00 a.m.
Speaker: Debbie Friedman, Director, MCH Trauma Programs
More info: Patricia Sidhom at 514-412-4400 x22599

Pediatric Courses

PEDIATRIC ADVANCED LIFE SUPPORT COURSE (P.A.L.S.)
(course offered in English; offered in French by request)
The P.A.L.S. course is fully accredited (13 credits)
Dates: September 27-28 or November 1-2
Description: The Montreal Children’s Hospital offers its Pediatric Advanced Life Support course (P.A.L.S) four times per year in September, November, January and April.
More info: Filomena Gonçalves, P.A.L.S. Administrator: filomena.goncalves@muhc.mcgill.ca
514-412-4400 ext. 23189
or Dr. Laurie Plotnick: drplotnick@videotron.ca

For further information and to register for these or any other MCH continuing education events available by teleconference, visit www.reseaudesanteenfant.ca and select <Continuing Education>.