

the Beat

SUMMER 2007, VOL. 15, ISSUE 1

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Transition Between Two Worlds

by Nathalie Comtois, RN

I recently attended a conference about Bridging the Gap Between Pediatric and Adult Care. This got me thinking about our clinic at the Montreal General Hospital and the challenges that occur with our youngest patients. It got me thinking about what we could do to ease the transition between both worlds, because it is two different worlds.

No drawings on the walls, no TV...and on top of this there are new doctors, nurses and surroundings. Except for the first visit, these young patients are coming to clinic by themselves. Here there is no talk about being fixed; we talk about another intervention or surgery. Not news to cheer them up and have them wanting to come back!

You must also take into account that it is much easier to miss school while in elementary or high school than it is to miss school in college or university, and often more difficult to miss work particularly if you have just started a new job.

How can we help? There are no miracle

answers. What we are trying to do is have all investigation and visits (ECG, echo, clinic visit) in one day that is convenient to each individual. We try to assist by avoiding exam periods etc. We have good communication with the Children's Hospital and other locations that refer to us, so that no one is left in the shuffle. We must remember that no matter what we do, the bulk of stress is on the patient. They are adjusting to adult life, the responsibilities that come with this, and a brand new set of health care professionals.

They also need to understand the importance of "wasting" a couple of hours with us once in a while to ensure that everything is OK. For these patients it is important for proper follow-up care by a specialist as it is often around late teen – early adult life that other interventions may arise. This conference helped open my eyes to the importance of talking about transition with the young patients and their families so, when the time comes for intervention or surgery, the news will not come as a surprise to anyone.

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A Newsletter

written by

Volunteers for

the well-being

of Adults

with Congenital

Heart Conditions

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Eisenmenger's Syndrome

by Doreen Fofonoff, MN, RN, CCN(C)

Eisenmenger's syndrome (ES) is a condition that includes pulmonary hypertension (high pressures in the lung arteries) and a shunt (flow) of blood from the right side of the heart to the left side of the heart.

How does ES develop?

People with ES are usually born with a large hole in the heart, such as ventricular septal defect, atrial septal defect, patent ductus arteriosus or a hole between major blood vessels, such as an aortopulmonary window. The hole in the heart causes blood from the left side of the heart (oxygen rich blood) to shunt (flow) over to the right side of the heart (oxygen poor blood). This causes oxygen poor and oxygen rich blood to mix on the right side of the heart. As a result, more blood than usual flows into the lung circulation. This causes the pressure in the lung circulation to rise. Over time, the high pressure damages the lung blood vessels. They may become thick, stiff and may even become blocked.

As the pressure in the lungs increase, the right ventricle (lower chamber of the heart) has a harder time pumping blood forward to the lungs. This may cause the right ventricle to enlarge. The high lung pressures also cause the shunt to reverse. Blood now shunts from right heart to the left heart, instead of from left to right. This causes oxygen poor blood to mix with oxygen rich blood on the left side of the heart. As a result, the blood pumped out to the body has lower levels of oxygen than normal. This is a condition known as hypoxia. The body's response to hypoxia is to produce more red blood cells so more oxygen can be carried.

How is ES diagnosed?

The cardiologist will take a medical history and do a physical exam to look for signs of ES. By listening to the heart with a stethoscope, the doctor can usually hear the swish of blood as it goes

through the hole in the heart.

Tests done to verify ES include:

- ECG to look for abnormalities in the heart's electrical system
- Echocardiogram, an ultrasound picture to look at the heart's structure, flow of blood and pressures
- Chest x-ray to look at the heart and lungs for enlargement or fluid
- Blood tests to do a complete blood cell count and measure kidney function
- Pulse oximetry to measure oxygen levels in the blood

What are the signs and symptoms of ES?

People with ES are cyanotic (bluish color to the skin, especially lips, fingertips and toes), have clubbed fingers or toes, increased red blood cells (erythrocytosis) and high hemoglobin. They may feel tired and breathless with exercise. They may also have irregular heart rhythms, chest pain, fainting or cough up blood (hemoptysis).

What health problems can occur with ES?

Stroke: People with ES are at increased risk of stroke. One reason is that the hole causing the right to left shunt of blood can allow air bubbles, bacteria or small clots to flow to the left side of the heart. From the left heart, they can go directly to the brain, block a blood vessel and cause a stroke. Usually, the lungs trap the bubbles, bacteria or clots. Air filters should be used on any intravenous (IVs) to trap the bubbles, bacteria or clots.

The second reason, patients with ES are at increased risk is because of the increased number of red blood cells. The large number of red blood cells may cause the blood to be viscous (thicker) which may increase the risk of clots forming.

Bleeding problems: The low oxygen levels in the blood result in decreased clotting factors and platelets (blood cells that help the blood to clot). This may increase the risk of sudden bleeding. Medications that affect the clotting of blood should be monitored closely.

Low iron levels: One of the most common causes of low iron in patients with ES is too frequent phlebotomies (removing blood). A low level of iron decreases the amount of oxygen that the red blood cells can carry. As a result, the person may not be able to exercise as much and may feel tired. Low levels of iron may also cause the red blood cells to become stiff and rigid. The red blood cells cannot then wiggle through the tiny arteries and a blood clot may form. This may increase the risk of stroke.

Abnormal heart rhythms: Abnormal heart rhythms such as atrial flutter, atrial fibrillation, ventricular tachycardia or ventricular fibrillation may occur with low oxygen levels or enlargement of the right side of the heart. Symptoms of irregular heart rhythms (palpitations), dizziness or fainting should be reported promptly to your doctor.

Coughing Up Blood: Coughing up blood (hemoptysis) is related to the high pressures in the lung, abnormal lung blood vessels and decreased clotting factors. Coughing up blood could be related to a lung blood clot, bleeding problem, or lung infection and should be reported immediately to your doctor.

Poor Kidney Function: Low oxygen levels may cause the kidneys not to work properly. Patients with ES may get gout because the kidney cannot clear waste products well from the blood. Certain medications, dyes used for heart tests, and dehydration can damage kidney function.

If I have ES, can I have children?

Women with ES should avoid pregnancy because of the increased risk of death and complications, both for the mother and the baby. Any pregnancy should be reported immediately to your congenital cardiologist. The cardiologist may recommend termination of the pregnancy. If the woman desires to continue a pregnancy despite being informed of the extreme risks then very close monitoring by a cardiologist, obstetrician and anesthetist familiar with

ES is recommended.

How is ES Treated?

The goal of treatment for patients with ES is to control symptoms and avoid problems. A cardiologist, with expertise in adult congenital heart disease, should see a person with ES one to two times per year. Some people may require more frequent check ups. The cardiologist will perform a thorough medical history and physical exam.

A therapeutic phlebotomy (removing a large amount of blood) may be done to remove excess red blood cells, thereby decreasing the hemoglobin. Phlebotomies may be recommended if the person has hyperviscosity symptoms (headache, dizziness, altered alertness, double or blurred vision, tingling of the fingers, toes, lips, fatigue, muscle weakness) or may be done before certain surgeries. Phlebotomies are usually not done more than three times per year. Too frequent phlebotomies may cause iron deficiency anemia and increase the risk of stroke.

Medications may be used to treat a variety of problems such as low iron levels, irregular heart rhythms, heart failure, high blood pressure, blood clots, and to improve the contraction of the heart muscle or to dilate (open) lung arteries. Patients with ES require close monitoring of their medications as any rapid change in blood pressure, fluid volume or heart rate could cause more right to left shunting of blood and further decrease oxygen levels.

Heart surgery to correct the hole in the heart cannot be done once ES has developed. The repaired heart is unable to push blood forward against the high pressures of the lung and will enlarge and fail. A heart and lung transplant or lung transplant with repair of the congenital defect may be an option for some patients.

What should I know about living with ES?

Know your heart: Learn the medical name of ES and your congenital defects so you can provide accurate information to anyone who provides care to you. Get a medic alert bracelet or necklace that lists ES and your congenital defects. The medic alert information will let health care providers know what your medical condition is so they can provide the best care for you in the event of an emergency. Ask questions of your health care team until you understand ES and how to care for yourself.

Get expert care for your heart: ES and your heart defects are very different than the usual adult heart problems. You should receive care from a cardiologist with expertise in ES and congenital heart defects. This will ensure that you receive the right treatment and care. See your congenital cardiologist regularly, even if you feel fine. Heart problems can start without symptoms. Early treatment can avoid more serious problems. Report any pregnancy immediately. Tell any new doctor, or dentist that you have ES. Let the laboratory know that you have ES and high hemoglobin. They will need to make certain adjustments in the blood collecting tubes.

Consult with your congenital cardiologist before having non-heart related surgery or procedures. Even a simple surgery requiring an anesthetic, such as a gall bladder operation, may be life threatening for a patient with ES. Surgery should be arranged with the congenital cardiologists, the surgeon, and a cardiac anesthetist. The surgery should be done at a hospital where the doctors have expertise in dealing with patients with ES.

Prevent Health Problems: Report to your cardiologist any increase in shortness of breath or feeling tired, irregular heart rhythms, chest pain, fainting, coughing up of blood or other bleeding, or symptoms of stroke (sudden numbness or weakness of the

face, arm or leg, especially on one side of the body, sudden confusion, trouble speaking or understanding, sudden trouble seeing in one or both eyes, sudden trouble walking, dizziness, loss of balance or coordination, sudden, severe headache with no known cause). Tell your cardiologist about any visits to emergency departments and admissions to hospital. Any health problem may affect the delicate balance of ES. obstetrician and anesthetist familiar with ES is recommended.

Prevention of pregnancy should be discussed with your cardiologist. Some methods of birth control may place women with ES at increased risk of clots, stroke and endocarditis.

Check with your cardiologist before taking any over the counter medications and herbal medications. Some medications may affect blood pressure, blood clotting or kidney function.

Avoid phlebotomies unless recommended by your congenital cardiologist. Avoid fasting, dehydration, and extremes of temperature.

Consult with your congenital cardiologist or clinic before traveling. Air travel is usually well tolerated but you may need oxygen on the flight. You should organize your trip well in advance to reduce emotional stress and physical exertion. Avoid dehydration. Make sure you have extra medical insurance when traveling outside of Canada as some provincial medical plans may not cover all costs should you become ill.

Prevent endocarditis: Endocarditis is a heart infection that happens when bacteria enters the blood and settles in the heart. It is serious and needs prompt treatment. See your doctor, if you develop a fever for which there is no obvious cause. Make sure the doctor takes a blood sample to identify the infection before you start any antibiotics. Prevent endocarditis by taking good care of your teeth, gums, nails and skin.

Take antibiotics as directed by your cardiologist before any dental or medical procedure.

Exercise Regularly: People with ES can usually perform most activities of daily living, but easily become tired. They should continue to keep active by doing light exercise as tolerated, avoiding extreme exertion (check with your cardiologist as to what is safe). Activities that cause extreme shortness of breath, light-headedness or fainting should be avoided.

Stay Healthy: Eat a heart healthy diet, rich in fruits, vegetables, whole grains and low in saturated fats and salt. Maintain a healthy weight and consult your cardiologist before starting any weight loss diet. Do not smoke, abuse drugs or drink alcohol to excess. Get regular flu shots.

CCHA Celebrates 3 Years

by Shelagh Ross, Vice-president and Communications Committee Chair

The Canadian Congenital Heart Alliance (CCHA) celebrated its third birthday this spring!

Three years ago, before leaving Toronto for the US, Dr. Gary Webb gave us some alarming “news”. Adult CHD care was in crisis and our future care was in jeopardy. I say “news” because it was a concept few of us had ever considered.

We found out that while the number of adult patients is now greater than ever due to excellent pediatric medical care, the provision of care for adults is minimal at best. Adult patients are dying unnecessarily due to a shortage of specialists trained in adult CHD, and unacceptably long waiting times for surgery and other interventions (33 times longer than for patients with coronary heart disease!). Incentives are lacking to attract doctors to specialize in Canada and we are more or less invisible to hospital budget-makers, the government, Heart and Stroke, and

other potential sources of support and funding. After being treated with kid gloves as children, the survivors are being ditched!

In response, a group of us banded together to form an alliance of CHD patients and family members to raise the profile of CHD in Canada. I'm happy to report that we've come a long way since that first meeting. We haven't exactly solved the problems, but we've taken steps to help patients while making noise at the same time. We're the first congenital heart defect group in Canada to bridge the gap between pediatric and adult patients, and our mission is to improve the lives of all CHD patients – *for life!*

We recently announced the first ever insurance package to members 40 years and over, which is quite a coup considering this will be a first for many of us. The package options include “no-medical” Life, and Health & Dental insurance. It will be available to our paying members in the coming months. As well, the CardiacKey is in a one-year pilot project right now at the Toronto Clinic and once complete will be available to all CCHA members.

We have also partnered with the Heart2Heart Cardiac Camp – the first teen CHD camp of its kind in Canada – which will be running Labour Day weekend in Ontario and Quebec. It will be a wonderful opportunity for teen CHD patients to learn more about their heart defects, meet adults with CHD, and have fun at the same time.

Our group has representation on a Special Advisory Panel convened by the Ontario Ministry of Health which is examining the state of care for Adult Congenital Heart Disease. We welcome the opportunity to have a voice at this forum. We have made strong ties with the American Congenital Heart Association and plan to join forces with more CHD organizations worldwide. There is also interest in starting CCHA chapters in BC and Quebec.

Last but not least, we are working hard to solicit donations to help develop new projects. If you can help, or know of someone who might be interested, we would love to hear from you. We have a lot of work to do!

For more information about us and what we have to offer, please visit us at www.cchaonline.ca or email us at info@cchaonline.ca.

For information about Heart2Heart Cardiac Camp, visit www.heart2heartcardiaccamp.ca.

Clinic Updates

Pacific Adult Congenital Heart Clinic

Our clinic continues to grow and expand and is exceeding the resources we have. We have submitted a proposal to government for more funding for physicians, nurse practitioners, a secretary, psychologist and social worker. We hope to hear soon whether funding is approved.

Suzanne Scott, one of our patients, is leading the initiative to establish a BC Chapter of the Canadian Congenital Heart Alliance (CCHA). The CCHA was established to raise the profile of adult congenital heart disease in Canada. Anyone wanting to get involved in this initiative is asked to contact Suzanne at suzepaul@hotmail.com

We are sad to announce that Wynne Chiu, our patient educator, left our clinic. She returned to school to complete both a Critical Care Nursing course and her Master's of Nursing degree. We thank her for all her support to the clinic and wish her well in her endeavours. We welcome to our team, Dr. Jennifer Ellis, a radiologist with an interest in cardiac MRI.

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Northern Alberta Adult Congenital Heart (NAACH) Clinic

The Northern Alberta Adult Congenital Heart program continues to be busy. Dr. Jeffery Smallhorn joined the Division of Pediatric Cardiology at the University of Alberta as director of the Pediatric Echocardiography and has introduced three-dimensional echocardiography. Many adult congenital heart patients benefit from this technology. Our surgeons find 3D-echo quite useful in planning for surgery, and we are also using it as a research tool to improve our understanding of congenital heart disease.

The Western Canadian Children's Heart Network, an organization dedicated to providing access to excellent cardiac care for pediatric heart patients, has expressed interest in expanding their mandate to include adult congenital heart patients. The WCCHN continues to sponsor a bimonthly adult congenital heart video teleconference for cardiologists and cardiac surgeons in Vancouver, Calgary, Edmonton, Saskatoon, Regina and Winnipeg. This facilitates referral for congenital heart surgery in Edmonton, and provides an opportunity for specialists to discuss complex cases and improve the care provided to these patients. The WCCHN has recently implemented a western Canada-wide database to which our site is contributing adult data. This will prove useful in the future to track outcomes of congenital heart patients, and it is hoped that other centres in western Canada will contribute to the database in the future.

We continue to develop expertise in catheter procedures in collaboration with our Pediatric Cardiology colleagues. Since last summer, 11 pulmonary valve replacements have been completed by catheter, replacing open-heart surgery for these patients. These valves are placed in previous homograft conduits and biologic prostheses. This is in addition to our usual volumes of atrial septal interventions (40-50/year) as well as other assorted interventions including coarctation stenting and PDA closures.

Toronto Congenital Cardiac Centre for Adults (TCCCA)

It has been a busy year in the Toronto clinic this year. With the ongoing addition of cardiologist, anesthesiologists, rhythm specialists, MRI specialist and fellows to our team we are creating quite a well-developed program. The clinic is also growing and we are pleased to report that we registered our 10,000th patient recently.

The Patient Conference was held May 12, 2007 and was yet again a success. There were many interesting speakers from all over the globe who presented diverse topics that built on the fantastic program from last year.

How you can become a CACH Member

To become a member of the Canadian Adult Congenital Heart Network you must complete an online form to be considered.

Visit this link for details:
www.cachnet.org/mem_app.html