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ALL THE POSSIBILITIES OF MODERN MEDICINE

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IN THIS ISSUE

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Prenatally Diagnosed Congenital Lung Lesions

Early diagnosis and treatment may prevent lifelong symptoms

Advances in medical technology are enabling physicians to detect problems at earlier and earlier stages of fetal development. Abnormalities of the developing lungs can be detected as early as four months gestation, enabling babies to receive treatment well before debilitating or dangerous symptoms develop.

In approximately one out of 30,000 pregnancies, a problem during development of the respiratory system leads to a birth defect in the lungs. This defect may manifest as one of three kinds of lesions:

- Congenital cystic adenomatoid malformations, or CCAM, cysts that replace some of the normal lung tissue;
- Bronchopulmonary sequestration, or BPS, in which abnormal lung tissue forms, and is nourished by extra blood vessels extending from the aorta; and
- "Hybrid" lesions, which may contain cysts and extra blood vessels.

Not all lung lesions result in symptoms, according to **Keith A. Kuenzler, MD,** *Director, Minimally Invasive Pediatric Surgery* at NewYork-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center (MSCHONY/CUMC). "It used to be that we would discover these lesions on X-rays or CT scans during the workup of children who had multiple bouts of pneumonia or difficulty breathing. Now, prenatal ultrasound has become so advanced that we find them long before any symptoms develop."

A percentage of prenatally detected lung lesions completely disappear on their own. In very rare cases, they may cause a fatal circulation problem in utero, known as hydrops fetalis. Some do not cause significant symptoms, but many do lead to recurrent lung infections, says Dr. Kuenzler. "These lesions take up space that the growing lung should occupy. While some are small, others may grow to take up half the chest." There is also an association between cystic lung lesions and unusual forms of lung cancer, although this risk is far lower than the risk of infection.

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The Agarwals' son had not yet developed respiratory symptoms from the hybrid lesion in his left lung, but Dr. Kuenzler performed a thoracoscopic left lower lobectomy in order to prevent problems such as shortness of breath, recurrent infections, or the rare possibility of malignancy.

The minimally invasive operation was performed June 12, 2009, and the boy returned home June 13 without needing pain medication. He was 'himself' June 14, according to his parents. At his postoperative visit June 29 (left), his incisions were healed and he had no pain. His tiny scars will likely fade completely.

Multi-Organ Autotransplantation

A new option for abdominal tumors once considered inoperable

In the past, surgical removal was not an option for some deeply embedded tumors involving the abdominal blood vessels. Now, transplant surgeons at NewYork-Presbyterian Hospital/Columbia University Medical Center (NYPH/Columbia) have developed a way to use a surgical method called autotransplantation to remove these otherwise inoperable tumors.

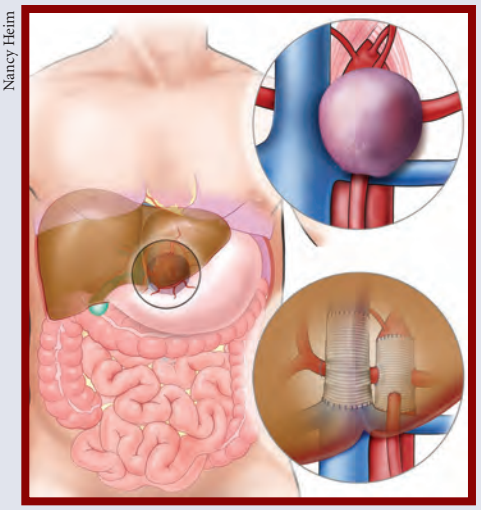
"Tumors involving the abdominal blood vessels have been considered inoperable because removing the tumor would cut the blood supply to vital organs," says **Tomoaki Kato, MD,** *Surgical Director, Liver and Gastrointestinal Transplantation.*

Autotransplantation is the removal and reimplantation of a patient's own organ (or organs). Unlike patients who receive organs from donors, patients undergoing autotransplantation do not have to wait for a donor to become available. They are not required to take immunosuppressant medications following surgery, as is the case with transplantation of donor organs.

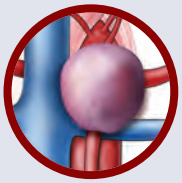
The state-of-the-art autotansplantation technique builds upon NYPH/Columbia's successes with multi-organ transplantation, most commonly used to treat patients with liver and intestinal failure. This year, Dr. Kato and a team of seven surgeons removed the small and large intestines, liver, pancreas, spleen and stomach from a seven-year-old girl. Three separate surgical teams worked to excise a tennis ball-sized tumor that was tangled among her vital abdominal organs and essential blood vessels. After removing the tumor, the team re-implanted the liver as well as the small and large intestines. The girl's pancreas, spleen and stomach – organs that had been compromised by the tumor – were unsuitable for re-implantation and could not be re-implanted. While the patient's life is no longer in danger, she will have some long-term medical challenges. Without her large intestine taking over the work of her stomach, she will have dietary restrictions comparable to that of someone who has had gastric bypass surgery. Without a pancreas, she will be diabetic. Absence of a spleen will possibly make her more susceptible to certain infections.

NewYork-Presbyterian Hospital is one of the few centers in the world with the expertise to perform multi-organ transplantation and autotransplantation. ■

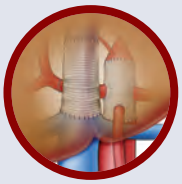
Nancy Heim



A lemon-sized tumor, attached to a patient's main arteries and wedged behind several organs, was considered inoperable by other institutions.



At NewYork-Presbyterian Hospital/Columbia, surgeons successfully removed the patient's liver, pancreas, spleen, stomach, small intestine, and two-thirds of the large intestine.



After excising the tumor safely, the team reconstructed the blood vessels and performed auto-transplantation to reattach the patient's organs.

Congenital Lung Lesions ~ continued from page 1

Obstetricians at MSCHONY's Center for Prenatal Pediatrics are often able to detect lung lesions at 15-25 weeks gestation. Except in the rare cases where dangerous circulation problems exist, babies with lung lesions are born normally, and after four months they undergo a CT scan. Lesions that persist are excised by removing the implicated lung lobe in its entirety. Surgery is performed between six and nine months of age, when the risks from anesthesia are reduced.

For the past two years, these operations have been performed at MSCHONY/Columbia with minimally invasive techniques, using three- and five-millimeter incisions and tiny instruments. This highly advanced technology allows the least tissue trauma, minimal scarring, and a very short recovery. Patients are discharged home the day following surgery, as opposed to the five days required for the larger, traditional open chest incision.

In Europe, physicians leave such lesions alone if a child is not experiencing any symptoms. In the U.S. however, these lesions are usually removed prophylactically. One important reason for this difference is reluctance to expose children to repeated CT. "We have the option of treating or observing such lung lesions, but following them requires repeated CT scans. Each CT scan produces the radiation equivalent of 100-150 X-rays, and no one knows the long-term effects of that much exposure," says Dr. Kuenzler. "It may also mean continuous anxiety for families over the course of years." As a result, most surgeons in the U.S. recommend surgical removal during the baby's first year. ■

Peripheral Vascular Disease

Cutting edge therapies and studies at NewYork-Presbyterian Hospital

As we get older, it is a natural thing to get pain in our legs when walking, right?

Wrong. Pain in the legs or feet while walking should not be considered a normal part of aging. It could be a sign of Peripheral Vascular Disease, or PVD – the buildup of fatty deposits (plaque) in the arteries beyond the heart. This buildup can block blood supply throughout the body, but most dangerously to the brain, kidneys, and legs.

Patients with even a moderate amount of arterial blockage may not have noticeable symptoms. Others may have symptoms, but not realize that their difficulty with walking is a result of PVD. Without the awareness of PVD, this problem may go undiagnosed, with patients suffering unnecessarily.

James F. McKinsey, MD, *Interim Bi-Campus Chief, Division of Vascular Surgery,* answers readers' questions about PVD.

Q: My doctor mentioned lower extremity vascular disease – is that different than PVD?

JFM: PVD may also be called lower extremity vascular disease or Peripheral Arterial Disease (PAD).

Q: What are the most common symptoms ?

JFM: Claudication (pain in the calf, thigh or buttock while walking), pain in the feet with elevation of the legs, and wounds or sores on the feet or legs that do not heal.

Q: How dangerous is PVD?

JFM: Even those with mild disease face an increased risk of heart attack and stroke. The leading cause of death in people with PVD is coronary artery disease (the buildup of plaque in the arteries of the heart), resulting in a heart attack. If PVD goes untreated, a patient's foot or leg may have to be amputated.

Q: I am overweight. Am I at risk for PVD?

JFM: Yes. PVD shares the same risk factors as heart disease – smoking, elevated cholesterol, diabetes, lack of exercise, and hypertension – which are frequently associated with being overweight or obese.

Q: How is PVD treated?

JFM: First, most patients need to make lifestyle changes such as quitting smoking, eating a healthier diet, and exercising more. Some people need medications to help lower cholesterol or blood pressure. In serious cases, patients can be treated with a minimally invasive procedure. A minority require surgery to clean out or bypass the arteries.

Risk Factors

The main risk factors for PVD are preventable: smoking, elevated cholesterol, diabetes, lack of exercise, and high blood pressure (hypertension). People with chronic kidney disease or diabetes who also smoke face an especially high risk of developing PVD. "Family history also plays a very important role in whether patients will develop vascular disease," says Dr. McKinsey. Although the specific genetic markers for PVD have not yet been identified, it is clear that certain populations (such as Hispanics and African Americans) have a higher predisposition to diabetes and PVD, and that some families have a genetic predisposition to developing PVD.

Correcting the preventable risk factors (through smoking cessation, regular exercise, and control of diabetes and hypertension) is very effective in treating PVD. "Those who don't do well are those who have a genetic predisposition and who don't do anything to reduce their risk," Dr. McKinsey says.

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Rajeev Dayal, MD, Assistant Professor of Clinical Surgery at Columbia University College of Physicians and Surgeons, screens a patient for PVD. He says, "Screening is a very easy and cost effective tool that can detect PVD, and potentially help to identify patients who have undiagnosed coronary artery disease."

Peripheral Vascular Disease ~ continued from page 3

Screening for PVD

Screening for PVD is simple, noninvasive, and cost-effective. It is done by measuring the blood pressure at the ankle with a blood pressure cuff and comparing this measurement with the blood pressure taken at the arm. Many cardiologists and vascular surgeons have this equipment in their offices today.

Nonetheless, screening is typically not reimbursed by insurance, and there is widespread debate about whether screening for PVD should be made a routine healthcare measure. Logic would tell us that it is important to be aggressive about identifying and controlling risk factors, but studies have not been done to scientifically prove that screening improves survival.

According to **Rajeev Dayal, MD**, Assistant Professor of Clinical Surgery at Columbia University College of Physicians and Surgeons, "Screening is a very easy and cost effective tool that can detect PVD, and potentially help to identify patients who have undiagnosed coronary artery disease." He and colleagues at NewYork-Presbyterian Hospital advocate that primary care physicians should regularly screen patients for PVD as part of routine checkups. Geriatricians in the U.S. also want to implement routine screening, but have faced resistance from insurers and challenges in purchasing the necessary machines.

Overall, most doctors recommend screening for people who smoke or have other risk factors. "If PVD is found early, it can be treated before extensive tissue loss occurs, potentially avoiding extensive surgical bypass procedures or even amputation," says Dr. Dayal. "When patients hear that they have PVD, it helps to convince some of them to stop smoking, lower their cholesterol, exercise more regularly, and make other lifestyle changes." ■

Research on PVD at NewYork-Presbyerian Hospital

The Division of Vascular Surgery is involved in a range of clinical trials on PVD. "Whether a patient is best served with the newest form of atherectomy, the latest design in bare metal stents or even gene therapy, we have it available within the Division of Vascular Surgery," says **Nicholas J. Morrissey, MD,** Director of Clinical Trials for the Division of Vascular Surgery.

In April 2008, Dr. McKinsey presented at the American Surgical Association the results of a study of almost 600 atherectomy procedures. This study at NYPH was the largest series with the longest followup ever published. Two thirds of the patients in the study had limb-threatening ischemia (would need an amputation if not treated). Over 94% of patients were able to salvage their legs and maintain good circulation three years after treatment, a result that is comparable or superior to outcomes achieved by open surgery.

Dr. McKinsey is the National Principal Investigator in the DEFINITIVE trial (Determination of EFfectiveness of the SilverHawk® PerIpheral Plaque ExcisioN System (SIlverHawk Device) for the Treatment of Infrainguinal VEssels / Lower Extremities), which aims to evaluate the effectiveness of the treatment of patients with all types of PVD who are treated by minimally invasive atherectomy.

Another important study at the Division of Vascular Surgery focuses on the treatment of patients with PVD and diabetes. "Diabetic plaque is very different than nondiabetic plaque," explains Dr. McKinsey. "Diabetics tend to have hard, calcified plaque, and this leads to a higher risk of gangrene. Patients with chronic renal failure have a similar type, and there is a relationship there. By evaluating all the treatment options – especially minimally invasive options as well as medical and gene therapy – we are hoping to identify the safest and most effective method to treat patients with PVD." ■

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