

Pediatric Views



Treating pediatric cataracts

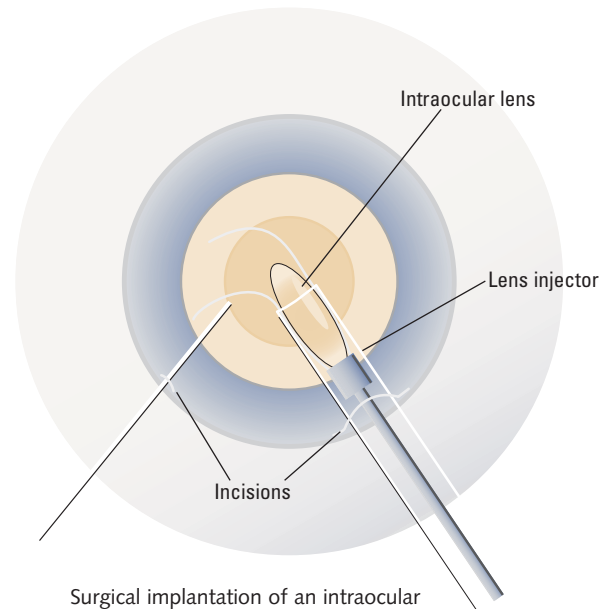


David Hunter, MD, PhD, Chair of Ophthalmology

Brennan Hughes-Shiverick was diagnosed with pediatric cataracts at 1 year old. He needed surgical removal of the cataracts, but with traditional methods, even after the procedure his parents would have needed to fit his tiny eyes with contact lenses for his vision to develop correctly.

Although mild cataracts in older children can sometimes be treated by patching the eye, using dilating eyedrops or prescription glasses, most pediatric cataracts require removal by surgery. Pediatric cataracts are uniquely challenging to ophthalmologists and are not like the cataracts that occur in adults. The eyes are too small for some of the standard cataract instruments. The lens itself can be extremely soft, rock hard or membrane-like, with increased risk of bleeding. The inflammatory response of the eye can be tremendous, requiring removal of the vitreous and intense steroid therapy after surgery. And after surgery, the child must be treated with amblyopia therapy, or the vision will not recover despite a good technical result.

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Surgical implantation of an intraocular lens. The folded lens is inserted through a 3 mm incision and unfolded to its full 13 mm size.

Early diagnosis, aggressive treatment benefit athletes



Lyle Micheli, MD, Director of Sports Medicine

Kevin Brake is an avid high school soccer and hockey player. When he began suffering from osteochondritis dissecans (OCD), a painful fragmentation of cartilage in his knee, his physicians initially advised him to take up the violin or guitar, because his sporting days were over.

Not ready to give up, Kevin's family turned to Children's Hospital Boston. **Lyle Micheli, MD**, director of Sports Medicine and a nationally recognized expert on sports injuries in children and adolescents, had a different assessment: with aggressive treatment, Kevin could be back on the field and in the rink within a few months.

The different prognoses, says Dr. Micheli, owe to the fact that the condition is rarely seen in most practices. "The average pediatric orthopaedist doesn't see a lot of OCD. Some see as few as one case per year, so understandably not everyone is up to speed with the effectiveness of new treatments. Here, we treat hundreds of OCD patients from all over the country every year."

OCD occurs when a loose piece of bone and cartilage separates from the end of the knee or elbow. That piece may stay in place or fall into the joint space, making the joint unstable. Patients usually present with pain, which is often poorly localized and associated with activity. Effusion may be present, and particularly in a case of an unstable lesion, the patient may describe mechanical symptoms such as locking or catching. If a loose body or bodies have developed, mechanical symptoms and episodic locking may predominate. Localization of symptoms depends to some extent on the location of the lesion.

While anyone may present with OCD, it occurs most often in males 10 to 20 years of age, while they are still growing. It affects athletes disproportionately, especially gymnasts and baseball players, and

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Q&A: Sudden death and sports

Charles Berul, MD



Charles Berul, MD
Pediatric Electrophysiologist, Associate in Cardiology

What causes sudden death during athletic activities?

The vast majority of sudden deaths are caused by cardiac events, with the remainder secondary to pulmonary causes such as asthma and anaphylaxis, or neurovascular congenital abnormalities such as aneurysms. Most sudden cardiac deaths in the young occur during or immediately following school sports, and nearly 90 percent occur in the presence of a teacher or coach.

How common is exercise-associated sudden death?

Among healthy children, sudden death while participating in sporting events is quite rare. Although uncommon, it attracts disproportionate attention from the media. In the United States, there are approximately 15 million school-age students and 100 to 200 reported pediatric sudden deaths per year, roughly calculating to one to two children per 100,000 annually. Sudden cardiac death during sports typically occurs in healthy, previously asymptomatic children and young adults.

Is it feasible or effective to screen healthy young athletes to detect those at risk?

The challenges of screening young athletes relate mainly to the difficulties in differentiating normal variations from true pathological markers of cardiac risk. Controversial issues include the accuracy of presymptomatic diagnostic testing and the cost-effectiveness of widespread pre-participation athletic screening evaluations.

How can care providers screen young athletes for risk factors?

The most effective means is evaluation by each child's primary care provider. Inquiring about pertinent symptoms will often be the most valuable diagnostic aid in identifying the rare, at-risk individual. Specific symptoms to inquire about include a history of syncope, chest pain, palpitations, dizziness or rapid heart rate. In particular, these symptoms become even more concerning when correlated with exercise. Candid discussions

are also necessary regarding legal and illicit drug use, alcohol, caffeine, smoking, and medications including prescription and over-the-counter drugs, health and nutritional supplements, and anabolic steroids. As some of the cardiac substrates for sudden death are

hereditary, a family history will also be consequential. A family history of congenital heart disease, arrhythmias, sudden death or inherited cardiac diseases (e.g., cardiomyopathies, long QT syndromes, Marfan syndrome) will markedly increase the level of suspicion. Such a family history justifies a more intense diagnostic assessment.

During the physical examination of a young athlete, the care provider should assess vital signs, symmetry of 4-extremity pulses, perfusion, weight and body habitus, and general overall health and fitness. Cardiac auscultation by the pediatrician or other primary care provider should include determination of the presence of murmurs, rubs, or clicks, and assuring a normal splitting and intensity of the S2 component of the heart sounds.

When do I need a subspecialist?

I recommend referral to a pediatric cardiologist if the primary evaluation findings include a concerning medical history, family history, physical or other potential risk factors, or in the case of exercise-associated symptoms or serious-sounding symptoms at rest.

What does a pediatric cardiologist/electrophysiologist do?

We have the task of determining whether the referred patient has an identifiable substrate for sudden cardiac death. He or she obtains a more organ-system-directed personal cardiovascular history and family history, and performs a cardiac-specific physical examination.

An ECG is relatively inexpensive and helpful for

CARDIAC/ELECTROPHYSIOLOGY TESTS

- ECG 12-lead, including manual measurement of intervals
- Chest X-ray
- Holter monitor, event recorder, or loop monitoring
- Echocardiography
- Magnetic resonance imaging
- Exercise tolerance test
- Cardiac catheterization
- Electrophysiology studies
- Tilt table testing
- Genetic screening tests

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How to reach us

Main number
(617) 355-6000

Call Center
(800) 355-7944

Emergency Services
(617) 355-6611

Transport Team
(866) 355-7944

TTY
(800) 355-8021

On the Web
www.childrenshospital.org

Sudden death and sports

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assessment of heart rate, rhythm, axis, conduction intervals, ischemia, infarction, and chamber size estimation. However, athletes may have ECG findings that overlap with pathologic abnormalities. In particular, a well-conditioned aerobic athlete may have sinus bradycardia, ectopic atrial or junctional escape rhythm, first- or second-degree atrioventricular block, intraventricular conduction abnormalities, premature atrial and ventricular contractions and/or exaggerated voltages suggestive of ventricular hypertrophy. These findings may be normal variants for the young athlete, and are sometimes difficult to differentiate from cardiac diseases.

A chest X-ray may reveal a generous-appearing cardiac silhouette, due to the relatively larger size of an athlete's heart. Imaging studies, such as echocardiography or magnetic resonance, are useful to assess intracardiac anatomy, chamber dimensions, and ventricular function. Exercise testing may be helpful for the correlation of exercise-associated symptoms, assessment of exercise intolerance, or for provocation (or suppression) of arrhythmias, conduction block, or exercise-induced ischemia. More invasive diagnostic studies are less frequently needed, and may be indicated based in part upon the history, physical examination and noninvasive evaluation.

Will pre-screening prevent sudden cardiac death?

Unfortunately, rare sporting catastrophes such as commotio cordis due to blunt chest wall impact cannot be anticipated by pre-participation screening. This is almost always fatal and occurs in normal children who are hit in the chest with an object (e.g., baseball, hockey puck or knee) during a vulnerable period of the cardiac action potential.

Are certain individuals and families more vulnerable to sudden death during sports?

There are patients with congenital heart diseases and cardiomyopathies that are particularly prone to development of arrhythmias and sudden death during exertion. These individuals can potentially be identified prior to sports participation, and should be given restriction guidelines or advised of specific exercise and sporting limitations. Inherited hypertrophic cardiomyopathy patients are especially vulnerable to exercise-associated sudden death, and this disease is the

leading cause of sudden cardiac death under age 35 in the United States.

There are multiple causes of QT prolongation on an ECG, including congenital and acquired long QT syndromes (see sidebar), but they can both result in sudden cardiac death during exercise. Typically, the congenital forms present during childhood, and the acquired forms may present during either childhood or adulthood, depending mainly on the inciting factors. Other electrical myopathies besides long QT syndrome, such as Brugada syndrome and arrhythmogenic right ventricular dysplasia, involve abnormalities in cardiac ion channel function, leading to ventricular arrhythmia vulnerability. Increased emotional and physical stress, as well as enhanced hormonal and catecholamine responses, associated with sports are likely triggers for the susceptible heart.

Where do we go from here?

Identification of at-risk individuals among a large population of young athletes is a challenge, particularly for primary care providers, who must attempt to identify patients with suspicious medical or family histories, or physical findings in order to selectively and accurately refer at-risk athletes for further subspecialty evaluation, diagnostic testing and possible activity restriction. The goal is to allow athletes full participation in activities without endangering their safety. This balance is precarious and requires careful attention to specific details and warning signs, such as exercise-associated symptomatology, and detection of familial cardiac diseases.

CAUSES OF PROLONGED QT INTERVAL

CONGENITAL

- Jervell & Lange-Neilson syndrome
- Romano-Ward syndrome
- Sporadic

ACQUIRED

- Electrolyte abnormalities
- Metabolic disturbances
- Malnutrition (especially anorexia nervosa, bulimia)
- Drug-induced
- Central nervous system trauma
- Ischemia, myocarditis
- Intraventricular conduction abnormalities

Who should people contact for more information or to refer a patient?

The Division of Electrophysiology within the Cardiology Department at Children's Hospital Boston has six physicians and four nurses who specialize in pediatric cardiac rhythm disorders. Outpatient programs are available in Boston and at satellite outreach clinics, including Lexington. You can also contact me directly at charles.berul@cardio.chboston.org.

To contact Children's electrophysiologists in Boston or Lexington, call (617) 355-6432. For more information on the Division of Electrophysiology, visit www.childrenshospital.org/electrophys.



Children's Hospital at Lexington

The state-of-the-art outpatient facility at Children's Hospital at Lexington offers 24 specialty services and 11 ancillary services, and is staffed by the same physicians who deliver world-class care at Children's Hospital Boston. With the recent addition of an outpatient electroencephalography (EEG) laboratory and an open extremities MRI, the Lexington facility now offers high-technology diagnostic tools once available only in the city.

Lexington opened its new EEG lab in January. It is staffed by Children's EEG technologists who perform the same diagnostic tests patients would undergo at Children's main campus. The recordings are electronically transmitted to Children's Department of Neurology in Boston for prompt interpretation by the epilepsy team.

Also available at the Lexington facility is a new, unenclosed extremity MRI. Patients with site-specific injuries sit comfortably in a chair, placing only their ankle, knee, hand, wrist or elbow into the doughnut-shaped magnet. The

open magnet is ideal for young children and patients made anxious or claustrophobic by a traditional MRI. Children's radiologists specializing in musculoskeletal injuries and disease read the scans the same day they are taken, making the entire MRI process quick and efficient.

EEG tests are available by appointment at 8:30 a.m., 10 a.m. and noon, every Tuesday and Thursday. Physicians can schedule a test by calling the clinic scheduling line at (781) 672-2100. Parents with specific questions or concerns about the process can call Children's EEG laboratory at (617) 355-7970.

For more information on the extremity MRI, contact Children's Department of Radiology in Boston at (617) 355-6286 or in Lexington at (781) 672-2200.

Children's Hospital at Lexington is conveniently located off of Rte. 128.

Pediatric and adolescent specialty referrals: phone (781) 672-2100, fax (781) 672-2145

Ambulatory Surgery: (781) 672-2300

Physical Therapy: (781) 672-2010

Radiology: (781) 672-2200

Laboratory: (781) 672-2030

SPECIALTIES AND SERVICES OFFERED AT LEXINGTON

Outpatient specialty services

- Adolescent Medicine
- Allergy
- Behavioral Medicine
- Cardiology
- Cerebral Palsy Clinic
- Dermatology
- Endocrinology
- Gastroenterology
- General Surgery
- Genetics
- Gynecology
- Hematology
- Lipid Clinic
- Neurology
- Ophthalmology
- Oral/Maxillofacial
- Orthopaedic Surgery
- Otolaryngology
- Plastic Surgery
- Pulmonary Medicine
- Rheumatology/ Immunology
- Sleep Disorders
- Sports Medicine
- Urology

Ancillary services

- Ambulatory Surgery Center: four state-of-the-art surgical suites
- Audiology: two sound booths
- Contact Lens Program
- Dermatology Laser Surgery
- EEG Laboratory
- Extremity MRI
- Nutrition Services: nutritional and dietary counseling
- Physical Therapy
- Phlebotomy
- Radiology: x-ray, fluoroscopy, ultrasound
- Speech Pathology

Inspiring future nurses

In March, Children's Hospital Boston's department of Nursing welcomed a group of high school students to the hospital to learn more about nursing careers. Part of Children's ongoing effort to address the statewide nursing shortage, the event gave students the opportunity to talk to nurses from a variety of specialties, tour the hospital and even watch a surgery in progress.



Cataract

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Brennan's problem was not the removal of the cataracts, but retraining his eyes to focus after the procedure. The options available to children his age are problematic. Contact lenses and prescription eyeglasses are an option, but pose serious compliance issues, especially in younger children. If a child fails to wear lenses regularly after the procedure, long-term outcomes will be affected.

The alternative is surgical implantation of intraocular lenses, but this has traditionally been avoided in children under 6 years because of their intense inflammatory response and high rate of implant rejection. However, under the leadership of **David Hunter, MD, PhD**, chair of Ophthalmology at Children's Hospital Boston, intraocular lenses are successfully being placed in younger and younger patients. "**Dr. Deborah VanderVeen** [assistant in Ophthalmology] and I have been successfully doing this procedure on children from 1 to 5 years old for long enough that it is now fairly routine for us, although it is never easy," says Dr. Hunter. "In some cases the procedure is appropriate for children as young as 6 months."

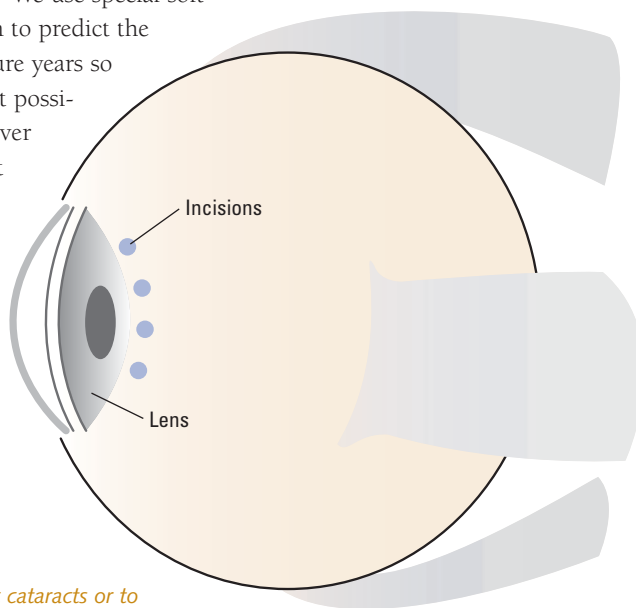
When Dr. Hunter met Brennan, he was certain surgery would be appropriate. He not only performed the procedure, but invited the child's ophthalmologist to participate. Dr. Hunter used a 3-mm knife to enter the anterior chamber of the eye. He aspirated the cloudy lens, then injected an acrylic lens into the existing lens capsule, where it unfolded to its full, 13-mm size. He removed the posterior capsule behind the implant to prevent clouding, as well as part of the vitreous to prevent formation of cloudy scar tissue. He closed the tiny incisions with ultra-fine, absorbable sutures.

"Choosing the lens power is a very complex process, since the eye is still growing," says Dr. Hunter. "We use special software in the operating room to predict the patients' eye growth in future years so that we can choose the best possible lens. Ideally, we will never have to replace the implant even though we know that the eye will grow."

The success of cases such as Brennan's owes both to the technology employed and the experience of Children's doctors, who see relatively high volume for a disease that affects just one in 2,000 children.

To learn more about pediatric cataracts or to contact Children's Department of Ophthalmology, call (617) 355-6401. Children's ophthalmologists also see patients in Lexington (781-672-2100) and Peabody (978-538-3600).

On April 18, Dr. Hunter will speak about pediatric vision screening at Caritas Good Samaritan Medical Center. For more information, call (508) 427-3547.



Small incisions are made at the edge of the eye's outer lining, or cornea, providing access to the anterior chamber.



Brazelton and Sparrow offer new parent resources

In a series of new books, famed Children's child development specialists **T. Berry Brazelton, MD**, and **Joshua Sparrow, MD**, spell out methods to help parents deal with some of the basic problems of childhood.

Sleep: The Brazelton Way covers everything a parent needs to know about sleep. Topics include the first great "touchpoint" of helping the baby sleep through the night; deciding whether to start with a "family bed" and how to wean a child into his or her own bed; sleepwalking; nightmares; and above all, helping children to go to sleep alone.

Calming Your Fussy Baby: The Brazelton Way helps parents interpret the very first cries of an infant, and shows them how to respond in the most appropriate and effective way. Drs. Brazelton and Sparrow deal with urgent problems such as colic, whining and tantrums. They also show how the emergence of gestures and words gradually replaces crying as the child's way to act on the world.

Discipline: The Brazelton Way shows how the normal physical, emotional and intellectual growth spurts can lead to conflicts and testing behavior. For each problem—defiance, lying, stealing, fighting, biting and foul language—Drs. Brazelton and Sparrow offer both understanding and practical solutions.

For more information, visit www.brazelton-institute.com.

A continuing commitment

New clinical documentation system to improve communication

Thanks to a new computer application rolled out last month, referring physicians will receive the most up-to-date clinical information on the patients they refer to Children's Hospital Boston in a more timely and reliable manner. The Web-based system known as Electronic Clinical Documentation (ECD) was designed by Children's Information Services Department to enhance functionality and replace a more archaic, character-based application called Electronic Signature (ESIG).

"The vast majority of referring physicians will receive clinical notes via fax at the time Children's attending physicians electronically sign them," says **Daniel Nigrin, MD, MS**, chief information officer. "Physicians in the community with admitting privileges to Children's who are currently able to edit and authenticate documents can also use ECD."

In addition to having a much more intuitive and stream-

lined graphical Web interface than the previous application, Children's physicians can view lab results and create documents such as operative notes, clinic notes and discharge summaries in the same system where they sign them. Other new functions include:

- Inserting portions of previous clinical documents into the document being edited.
- Creating, managing and inserting templates.
- Editing CC lists.
- Adding "sticky note" messages.
- Adding an addendum to an existing authenticated document.

If you have admitting privileges to Children's and have questions about using ECD, or are a referring provider looking for more information about the program, help is available at (617) 355-2002.

Save the date

Office Manager and Referral Coordinator Spring Meeting

May 8, 10:15 a.m. to 2 p.m.

Topics:

- An update on emergency services and transportation
- Injury prevention
- A review of coordinated specialty programs
- Tour of Children's Hospital Boston

For more information or to register, call (617) 355-2454.

Prenatal Cardiac Intervention Symposium

June 27 to 28 in the Enders Auditorium at Children's Hospital Boston

Topics include:

- Echocardiographic evaluation of fetal physiology
- Aortic stenosis and the evolution of HLHS
- Modifying congenital CHD in utero to improve post-natal outcome
- Access to the fetal heart
- Advancing the field of fetal surgery

For more information, visit www.cvp-chboston.org/pci or send an e-mail to pciinfo@cardio.chboston.org.

Celebrating a century of service

Wednesday, June 11 from noon to 1 p.m. at Children's Hospital Boston

In celebration of its 100th anniversary, the Children's Hospital Boston Alumni Association presents the 50th annual Blackfan Lecture. Presented by Lawrence H. Summers, President of Harvard University. For more information, call (617) 355-2291.

CME corner

Children's Hospital Boston and Harvard Medical School's Department of Continuing Education present the following Pediatric Health Care Summits. The summits are free, community-based continuing medical education seminars designed to inform primary pediatric providers of trends in the management of common pediatric health concerns. For more information or to register for one of the following courses, visit www.childrenshospital.org/resources/cme/courses.cfm or call Physician Relations at (617) 355-2454.

Date	Topics and Speakers	Location and Credits
May 20	<ul style="list-style-type: none"> • Imaging of Child Abuse Paul Kleinman, MD • Immunizations: What's New for 2003 Henry Bernstein, DO • STDs in Teenagers Lydia Shrier, MD • Common Problems in Pediatric Nephrology Michelle Baum, MD 	<p>Beverly Hospital Beverly, Mass. 4 hours</p>
June 19	<ul style="list-style-type: none"> • Fundamentals of Hypoglycemia: A Physiologic Approach David Weinstein, MD • Issues in Adoption Lisa Albers, MD, MPH • An Update on Lyme Disease Richard Malley, MD 	<p>South Shore Hospital Emerson Conference Room South Weymouth, Mass. 3 hours</p>

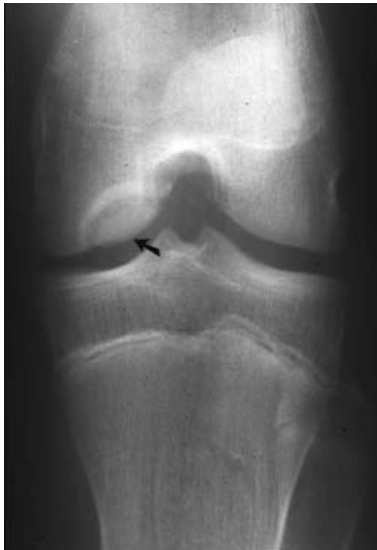
OCD

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has been increasingly seen in adolescent girls as that group has become more active in team sports. While the etiology of OCD is still debated, its apparent increased prevalence in the knees of children involved in organized sports suggests repetitive impact training as a major factor.

According to Dr. Micheli, the key to successful treatment is early diagnosis. “At least half of OCD

Cast or brace immobilization is a viable but relatively conservative strategy, often used initially in younger patients. Active adolescents such as Kevin Brake, for whom prolonged immobilization and activity restriction are unacceptable and have a lower rate of success, benefit from more aggressive approaches, such as transarticular drilling (which creates channels for revascularization and healing within the articular cartilage), antegrade debridement with replacement and fixation, or retrograde grafting and stabilization. Dr. Micheli estimates that about 70 percent of the



Large OCD of medial femoral condyle in a teenage boy.



Same lesion shortly after compression screw fixation.



Complete healing eight weeks later.

patients referred to us have experienced a delay in diagnosis,” he says. “In some cases that’s because the symptoms are common to other conditions, such as juvenile arthritis or Lyme disease. In other cases the patient is told to ‘give it time.’ ”

Dr. Micheli maintains that any sports-active child with unexplained knee pain should be thoroughly assessed. “If there is swelling or effusion,” he adds, “that should be a red flag.”

Another problem Dr. Micheli sees is that some care providers aren’t aware that in many circumstances OCD can be treated effectively. Effective treatment of OCD was pioneered at Children’s in 1951 by **William Green, MD**, who showed that an immobilized joint could heal properly. Today, Sport Medicine specialists at Children’s have a wide range of treatment options depending on the age and relative maturity of the patient, the size and location of the lesion, whether the lesion is open or closed and whether it is mechanically unstable or involves a loose body.

OCD patients his department sees are candidates for aggressive surgical treatment.

In Kevin’s case, Dr. Micheli applied transarticular compression screws to reconnect the articular cartilage to the bone. Just six months later, Kevin’s family sent a letter to Dr. Micheli reporting that he had finished the soccer season with no symptoms and had returned to ice hockey. “By all opinions,” they wrote, “Kevin is again a college prospect in soccer. Based on all our early consultations... we never thought we’d see this level of recovery.”

As far as Dr. Micheli is concerned, the negative prognosis Kevin Brake received is all too common for young athletes with OCD. “The bottom line,” he says, “is that if OCD is caught early enough and treated aggressively, the devastating outcomes we’ve traditionally seen can be avoided.”

For more information visit www.childrenshospital.org/sportsmed. To contact Sports Medicine or to refer a patient, call (617) 355-6028 (Boston) or (781) 672-2100 (Lexington).

New additions to the Dept. of Radiology:

Rick Fair, MD
Staff Radiologist
General Radiology

Susan Connolly, MD
Staff Radiologist
General and
Musculoskeletal
Radiology

Anna Golja, MD
Staff Neuroradiologist
Neuroradiology

Jeanne Chow, MD
Staff Radiologist
General Radiology,
Fetal Imaging and
Genitourinary Radiology

Jeannette Perez-Rossello, MD
Staff Radiologist
General Radiology

More information:
www.childrenshospital.org/radiology or call
(617) 355-6286

New addition to the Dept. of Ophthalmology:

Linda Dagi, MD
Director, Strabismus
To schedule an
appointment, call
(617) 355-6401 (Boston)
or (781) 672-2100
(Lexington)

Children's specialists are closer than you think



Spotlight Site

Children's Hospital at Lexington

Pediatric outpatient specialty services include:

- Adolescent Medicine
- Cardiology
- Gastroenterology
- Ophthalmology
- Orthopaedics
- Urology

For more information on Children's Hospital at Lexington, see page 4.

Children's physicians provide multi-disciplinary pediatric specialty care at locations throughout Eastern Massachusetts.

Children's specialists in cardiology, gastroenterology, neurology and other pediatric subspecialties are easily accessible to you and your patient families.

Locations are right off Rte. 3, Rte. 24, Rte. 128, I-93, I-95 and the Mass Pike.

For more information about the Children's Health Network site closest to you, or for more information about Children's Hospital at Lexington, please contact Children's Call Center at (800) 355-7944, or visit our Web site at www.childrenshospital.org.

Pediatric Views



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