

*A Special Place for*

# Pediatric Special Procedures

By Scott P. Edwards



Dr. Mary-Alice Abbott is able to treat children with a variety of genetic disorders on the Pediatric Special Procedures Unit at Baystate Children's Hospital.

The bright colors and wall-sized murals on the Pediatric Special Procedures Unit at Baystate Children's Hospital belie the fact that many of the children who come to the unit are very sick, often with rare diseases that cannot be treated elsewhere in this region.

The unit is designed to provide advanced care and treatment for Baystate's youngest patients away from the bustle of the Pediatric Intensive Care Unit (PICU), but with all of the benefits of an intensive care unit.

The unit was originally designed and developed by Anastasios Angelides, MD, chief of Pediatric Gastroenterology, and pediatric surgeon Barry Sachs, MD, about ten years ago for procedures requiring anesthesia support outside of the operating room. "It was a novel model we adopted that is now common in other children's hospitals," says Dr. Angelides.

"Having this special outpatient unit has many benefits, such as reducing potential exposures to other sick children for procedures or infusions that would otherwise be performed in the PICU," says Robert Kaslovsky, MD, chief of Pediatric Pulmonary Medicine. "It is exclusively a pediatric unit, run by a staff that is familiar with the needs of the children, as well as the clinicians working there."

Leslie Murphy, RN, CCRN, is the PICU manager and also oversees the Pediatric Special Procedures Unit, located on the second floor of Baystate's Springfield Building. She says the unit provides a range of services, including pediatric endoscopy and bronchoscopy, short chemotherapy infusions, lumbar punctures, bone marrow biopsies and aspirations, PICC line insertions for long-term IV antibiotics, and minor surgical procedures.

In addition, she says, Baystate has started a new Infusion Program on the unit for children with inflammatory bowel disease, juvenile rheumatoid arthritis, immune disorders, and rare genetic diseases, among other conditions.

"Besides providing special procedures and infusions," she says, "our team offers special, personalized care for children and their families. We are committed to providing a memorable health care experience – one

that addresses each family's unique needs and empowers them to be involved in their treatment. The unit is patient and family centered, as is all of Baystate Children's Hospital, and we welcome input from the patient and family regarding their plan of care."

The Infusion Program also **allows treatment** for rare genetic diseases like osteogenesis imperfecta and Pompe disease.

## Bronchoscopy and Endoscopy

Dr. Angelides notes that the Pediatric Special Procedures Unit is used most often for diagnosis and treatment of pediatric gastrointestinal disorders. Endoscopic procedures can be performed safely even in the smallest infants for conditions ranging from acid/peptic disorders and inflammatory bowel disease to malabsorption syndromes and abdominal pain.

Baystate's pediatric pulmonologists perform outpatient flexible bronchoscopy with deep sedation or general anesthesia on the Pediatric Special Procedures Unit. According to Dr. Kaslovsky, there are many indications for bronchoscopy in infants, children, and adolescents including: stridor, persistent wheezing, chronic cough, suspected foreign body aspiration, gastroesophageal reflux with pulmonary symptoms, airway evaluations in tracheotomy patients, and recurrent pneumonia. Bronchoscopy procedures can be coordinated with GI endoscopy under the same anesthesia.

"In many cases," says Dr. Kaslovsky, "flexible bronchoscopy may allow practitioners to reach a diagnosis when physical exams, spirometry, and x-rays are not

revealing.” He notes that flexible bronchoscopy can also be used therapeutically for children with pneumonia or complete or partial atelectasis.

## Infusion Program

The unit’s Infusion Program fills a niche in the region. While genetic diseases are rare, and thus the number of patients are relatively low, says Mary-Alice Abbott, MD, chief of Medical Genetics, “if you put them together, it’s a pretty good number of kids.”

In addition to treating more common problems such as irritable bowel syndrome, the program tackles rare genetic diseases like osteogenesis imperfecta, which is caused by faulty collagen and causes fragile bones, and Pompe disease, which is caused by a lack of or faulty enzyme needed to metabolize lipids.

Dr. Abbott treats area adults and children with Pompe disease, the rare genetic disorder featured in the Harrison Ford movie *Extraordinary Measures*, using enzyme replacement therapy (ENT). An inherited, often fatal disease, Pompe is caused by a mutation to an enzyme that breaks down glycogen. The mutation causes excessive amounts of the enzyme to build up in the body, choking off cells, in particular in the muscles and heart.

Left untreated, most children with Pompe die before their first birthday. In one case, however, Dr. Abbott initiated ERT in a baby girl less than two weeks old. The child is now three years old and while her life expectancy is shortened, Dr. Abbott says she has far exceeded expectations from when she first began therapy. “This unit,” she says, “means that patients like this little girl have regular access to life-saving infusions.”



Pediatric specialists, like Drs. Anastasios Angelides (left) and Robert Kaslovsky (right), perform a range of procedures on the unit, including pediatric endoscopy and bronchoscopy.

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## Regional Resource

The Pediatric Special Procedures Unit allows Baystate Children's Hospital to be one of the only places in the region offering many of these intensive therapies and treatments. "The unit allows us to treat children who would otherwise have to travel to Boston or elsewhere for procedures or infusions," says Ms. Murphy. "That can be a big trek. Some come every three weeks for treatment, so having this level of care available locally at Baystate Children's Hospital is more convenient and a big time-saver for their families."

The Pediatric Special Procedures Unit is open Monday through Friday from 8 am to 4 pm, with plans to expand to 16-hour days on weekdays and 8 hours on Saturdays, as well as the possibility of adding a seventh day to make access even more convenient for working parents and their children.

Unit manager Leslie Murphy, RN, notes that having this level of care available locally at Baystate Children's Hospital is more convenient and a big time-saver for families who would otherwise need to travel as far as Boston.

