In left ventricular outflow tract disease (LVOT), the topic of the 2018 Heart Summit, an obstruction affects the flow of blood from the heart.
Innovative therapies
Harnessing the newest insights and techniques for children’s care

BY THOMAS T. SATO, MD

As we begin a new year, we reflect on the highlights of the past year at Children’s. One of these was the Heart Summit, at which we welcomed pediatric cardiologists and cardiothoracic surgeons from institutions worldwide to share treatment and research discoveries. This group gathered to share as much evidence as possible, so that we all can learn what is working and what alterations in therapies can be most beneficial for our young patients. And, in the spirit of building medical consensus, we engaged in a spirited examination of various clinical approaches.

As we did at the first Heart Summit in 2017, attendees addressed the many aspects of congenital heart disease, and the fact that we now have a growing population of patients who underwent heart repair as infants and face new cardiac complications as they age into adulthood. This group still needs specialized care, and the Summit offered opportunities for practitioners to share innovative ideas on treatment and ongoing management.

Further innovation has come in the form of a remarkable immunotherapy treatment for a resistant type of leukemia. By altering a patient’s own T-cells, we are able to mount an immune response to attack cancerous blood cells. We are truly fortunate to have such incredible researchers and providers here at Children’s who offer this leading-edge treatment along with the most compassionate care.

Best,

Thomas T. Sato, MD, FACS, FAAP
CEO, Children’s Specialty Group
Senior Associate Dean of Clinical Affairs,
Professor of Pediatric General and Thoracic Surgery, Medical College of Wisconsin

A new way to treat childhood leukemia

Children’s is one of only about 40 pediatric hospitals in the country certified to offer cutting-edge CAR-T immunotherapy to children with relapsed or refractory B-cell acute lymphoblast leukemia (ALL). Pediatric Rounds interviewed Julie An M. Talano, MD, a pediatric hematologist-oncologist who leads the bone marrow transplant clinical research team, about this innovative therapy. Read the article at chw.org/car-t.
Meeting of minds
At the Herma Heart Institute’s annual Heart Summit, medical experts focused on a complex condition

This past October, Children’s Hospital of Wisconsin welcomed physicians from some of the world’s leading institutions to the Heart Summit, the Herma Heart Institute’s signature conference, where medical experts exchange ideas on a complex cardiac condition to help improve treatments and build medical consensus. This year’s topic was complex left ventricular outflow tract disease (LVOT).

LVOT is a rare defect — accounting for 5 to 10 percent of all congenital heart defects — in which an obstruction affects the flow of oxygen-rich blood from the heart. Numerous variables, including the exact nature and location of the obstruction, can affect the severity and treatment of the condition. The Heart Summit featured presentations on LVOT, as well as robust discussions and a live open-heart surgery.

INTERACTIVE LEARNING
The most anticipated component of the Heart Summit was the live surgery that Viktor Hraska, MD, PhD, surgical director of the Herma Heart Institute, performed on a 4-year-old child with LVOT.

Heart Summit participants followed Dr. Hraska’s procedure via a live, 3D video feed. The one-of-a-kind setup breaks down the wall of the OR and allows audience members to get an in-depth look at what happens during open heart surgery. Not only did attendees witness the surgery in breathtaking detail, but they were able to speak with Dr. Hraska and ask him questions.

“Our intent is to move out of the traditional classroom and one-way video streaming, and let an audience of cardiologists, surgeons, anesthesiologists, intensivists and the entire care team see inside the operating room — in 3D — to gain deeper understanding of what is actually happening during surgical repairs,” said Aaron Kinney, DBA, FACHE, executive director of the Herma Heart Institute. “This helps surgeons discuss techniques in real time, helps cardiologists better appreciate the before and after, and helps the rest of the care team increase their familiarity of specific surgical repairs.”

LASTING IMPACT
The Heart Summit’s interactive format allowed all in attendance to gain valuable insights on LVOT. This will ultimately help health care teams make the most educated decisions possible for their patients.
Pulmonary hypertension (PH) can present at any age from infancy to adulthood. In the pediatric population, it can be inherited, associated with other conditions such as congenital heart disease or chronic lung disease, or occur with no clear etiology.

The advent of targeted therapies in the past few years has dramatically improved the survival rate in children. As one of just eight pediatric PH centers in the United States to receive the Pulmonary Hypertension Association’s highest accreditation as a Center of Comprehensive Care, the PH program at the Herma Heart Institute sees some of the most complex cases in the country.

In a web exclusive article, Edward Kirkpatrick, DO, program director of pulmonary hypertension at Children’s, presents two case studies that demonstrate the PH program’s out-of-the-box thinking and multidisciplinary approach. Read “Innovative approaches to pediatric pulmonary hypertension,” by Dr. Kirkpatrick at chw.org/phpedsrounds.
Gender identity refers to a person’s internal sense of being male or female. A child’s sense of their own identity develops over time, and sometimes the match between a child’s assigned gender and their gender identity is not clear.

At Children’s Hospital of Wisconsin, we recognize the need for a health clinic devoted to children and adolescents who have concerns of any kind relating to gender identity. Our team is dedicated to providing care, education and support to patients and their families, while being sensitive to different experiences and circumstances.

**MULTIDISCIPLINARY CARE**
The Gender Health Clinic, located at Children’s in Milwaukee, is a multidisciplinary clinic focused on children and youth seeking assistance with gender identity development and transition concerns. Our specialists include:
- Pediatric endocrinologist
- Endocrinology nurse practitioner
- Pediatric health psychologist
- Endocrinology nurse

**WHAT WE DO**
Our skilled staff meet with patients on an individual basis for comprehensive evaluations, mental health consultations and education. We may provide puberty-suppressing hormone therapy or gender-affirming hormone therapy. We also provide patients and their families with resources and support within the community.

**A minute of your time**
Children’s Hospital of Wisconsin launched the first specialty-specific referring provider survey in May 2018, focusing on the Neurology department.

During this pilot project, when a provider refers a new patient to Neurology, they receive a short survey by email (maximum of one per month). The email arrives after the patient is seen by a Neurology provider, and it includes seven questions about the referral experience, the Children’s provider and Neurology services.

“We encourage referring providers to take the time to really let us know what they’re thinking,” said Carey A. Ehlertr, MD, a neonatologist at Children’s, associate professor of Neonatology at the Medical College of Wisconsin, and director of provider engagement and experience for Children’s Specialty Group.

Responses are shared with the department and individual providers. “It gives both the provider and the specialty actionable feedback,” Dr. Ehlertr said.

**WHAT’S NEXT?**
In the fall, Children’s launched a Pediatric Cardiology survey focusing on services in the Fox Valley region. Children’s plans to launch a survey on another specialty this spring.

**BY THE NUMBERS**
Our pilot survey has generated a good response rate. Here is a snapshot of results so far.

Rating Children’s Hospital of Wisconsin Neurology services on the following areas:

- The ease of the referral process
  - Excellent: 45%
  - Very Good: 30%

- The timely communication and reporting of patient information back to you by the provider
  - Excellent: 52%
  - Very Good: 24%

- The provider treating you as a valued member of the care team
  - Excellent: 52%
  - Very Good: 23%

**Contact the Gender Health Clinic at**
(414) 607-5280 or (877) 607-5280.
To refer a patient, call (800) 266-0366.
Tongue tie in infancy

Cutting through the controversy regarding treatment and classification of ankyloglossia

BY SOPHIE G. SHAY, MD

Ankyloglossia, commonly known as “tongue tie,” refers to congenital oral anomalies with varying degrees of restricted tongue mobility. It typically involves an abnormally short or thick lingual frenulum (the tongue web), or a highly attached genioglossus muscle, which tethers the tip of the tongue, limiting tongue motion or function.1–4

Sophie G. Shay, MD, is a board-certified pediatric otolaryngologist at Children’s Hospital of Wisconsin and an assistant professor of pediatric otolaryngology at the Medical College of Wisconsin.
Long-term complications from untreated ankyloglossia may include inadvertent biting of the frenulum, difficulty licking lips and socially debilitating problems with speech articulation. However, there is significant controversy regarding the treatment or even classification of ankyloglossia, including its impact on feeding and speech.

**ETIOLOGY**
The etiology of ankyloglossia is generally unknown. Most infants are born healthy with no other congenital abnormalities. A few case reports suggest an association between ankyloglossia and rare congenital syndromes such as X-linked cleft palate syndrome, Kindler syndrome, van der Woude syndrome, and Opitz syndrome.

Several studies have suggested a genetic basis for ankyloglossia, with positive family histories in 10 to 53 percent of patients.

Specifically, mutations of the TBX22 gene have been found in patients with cleft palate and ankyloglossia. Maternal cocaine use during pregnancy has also been associated with ankyloglossia, with a potential 3.2-fold higher incidence among children exposed to cocaine in utero.

**DIAGNOSING ANKYLOGLOSSIA**
Diagnosing ankyloglossia is particularly challenging given that there is no universally accepted classification system. It is also difficult to assess functional limitations in newborns because neither the maternal nipple nor the infant’s tongue is visible during breastfeeding.

Thus, diagnosis should be made after a careful, examination of the undersurface of the tongue and frenulum, as well as a thorough functional assessment.

- Anterior ankyloglossia may be easier to diagnose given the prominence of a shortened

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**Incidence of ankyloglossia**

- Overall incidence of ankyloglossia: 10.7
- Ankyloglossia among oral mucosa abnormalities: 0.1
- Ankyloglossia alone: 10.7
- 4.4
- 4.2

The incidence of ankyloglossia varies from 0.1 to 10.7 percent, depending on the population studied.

Studies on oral mucosa abnormalities find a 0.1-4.4 percent prevalence of tongue tie, while studies evaluating ankyloglossia alone report prevalence rates between 4.2 and 10.7 percent.
frenulum and tethering at the tip (Type I) or slightly behind the tip (Type II) of the tongue.

- Posterior ankyloglossia is more diagnostically challenging given its subtle physical findings and functional impairment. It is typically characterized by a thickened frenulum (Type III) or a submucosal frenulum presenting as a flat, broad mound absent of any typical protruding frenular tissue, and restricting base of the tongue mobility (type IV).³,²³,²⁴

**TREATMENT OPTIONS**

The decision to treat ankyloglossia, which treatment to use, and even the long-term effectiveness of treatment remain controversial.

Two surgical options are available: frenotomy (also called frenulotomy) and frenuloplasty.

**Frenotomy** is a quick, relatively simple procedure that can correct both anterior and posterior ankyloglossia. The procedure is generally performed in the office early in infancy for infants with feeding difficulty.²⁵ Older infants and children may require general anesthesia. The tongue is superiorly retracted to expose the lingual frenulum, which is then sharply incised, usually with sterile iris scissors. It is important that the clinician avoid the sublingual and submandibular gland openings along the floor of mouth by cutting the frenulum close to the ventral surface of the tongue. No suture is required, and there is typically very little blood loss. The infant may feed immediately following the procedure.

The most frequently reported complication is minor bleeding, which can be resolved with local pressure. There are rare reports of ulceration at the surgical site and delayed wound healing, injury to the submandibular ducts, surgical site infection, and sublingual hematoma.²⁵,²⁶

Who should perform frenotomy?

Frenotomy has been shown to be a largely safe procedure, with the majority experiencing no complications.²⁶,²⁷ Nonetheless, in order to minimize risk, frenotomy should only be performed by experienced and trained practitioners.
What is Z-plasty?

An alternative method of frenuloplasty is a Z-plasty technique, which releases the lingual frenulum and increases the length of the ventral tongue scar. In a classic Z-plasty, opposing 60-degree triangles are created and rotated to lengthen the scar. The lingual mucosa is then closed with absorbable sutures.

Studies comparing frenotomy to Z-plasty frenuloplasty find the latter provides significantly greater improvements in speech articulation, increased lingual mobility, frenulum length, tongue protrusion and parent satisfaction.
### Table 1. Common grading systems for ankyloglossia

<table>
<thead>
<tr>
<th>Grading system (source)</th>
<th>Anatomical classification criteria</th>
<th>Posterior ankyloglossia classification</th>
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</table>
| Cryllos system (American Academy of Pediatrics Section on Breastfeeding, 2004) | Type 1: Attachment of the frenulum to the tongue tip, usually in front of the alveolar ridge  
Type 2: 2–4 mm behind the tongue tip and on or just behind the alveolar ridge  
Type 3: Attachment to the midtongue and middle of the floor of mouth  
Type 4: Against the base of the tongue | Consists of type 3 and 4 with functional impairment |
| Kotlow system (Kotlow, 1999) | Normal: >16-mm free tongue length  
Class I (mild): 12 to 16-mm free tongue length  
Class II (moderate): 8 to 11-mm free tongue length  
Class III (severe): 3 to 7-mm free tongue length  
Class IV (complete): <3-mm free tongue length | Consists of normal and class I with functional impairment |
| Kotlow system revised (Kotlow, 2011) | Class I: 0 to 3-mm attachment from the tongue tip  
Class II: 4 to 6-mm attachment from the tongue tip  
Class III: 7 to 9-mm attachment from the tongue tip  
Class IV: 10 to 12-mm or submucosal attachment from the tongue tip | Consists of classes III and IV with functional impairment |
Mild: 17–22 mm  
Moderate: 4-16 mm  
Severe: 3 mm | NA |
| Tongue protrusion (Lalakea and Messner, 2003; Messner and Lalakea, 2002) | Normal: 20–25 mm  
Ankyloglossia: <15 mm | NA |

**Ankyloglossia and breastfeeding**

Breastfeeding challenges as a result of ankyloglossia include difficulty with latching, maternal nipple pain, and insufficient feeding, which may put the infant at risk for early infant weaning or failure to thrive in more extreme cases.\(^5\)

However, there is insufficient evidence to clearly associate ankyloglossia with difficulties in breastfeeding due to large variations in diagnosis of ankyloglossia and outcomes measures.

Nonetheless, several studies suggest that surgical intervention through frenotomy or frenuloplasty may improve breastfeeding efficacy, although the evidence is mixed.\(^35\)
There is currently insufficient evidence to clearly associate ankyloglossia with difficulties in breastfeeding or speech due to large variations in the diagnosis of ankyloglossia and outcomes measures. However, there are a number of studies that suggest surgical intervention through frenotomy is beneficial in improving breastfeeding efficacy. Surgical intervention in infants should be recommended with caution, typically in those with ankyloglossia clearly evident on physical examination and who have documented difficulties with breastfeeding. Procedures to treat ankyloglossia should only be performed by trained, experienced providers.

CONCLUSION

There is currently insufficient evidence to clearly associate ankyloglossia with difficulties in breastfeeding or speech due to large variations in the diagnosis of ankyloglossia and outcomes measures. However, there are a number of studies that suggest surgical intervention through frenotomy is beneficial in improving breastfeeding efficacy. Surgical intervention in infants should be recommended with caution, typically in those with ankyloglossia clearly evident on physical examination and who have documented difficulties with breastfeeding. Procedures to treat ankyloglossia should only be performed by trained, experienced providers.

REFERENCES


Keisha Adams, MD, is an adolescent medicine specialist at Children’s Hospital of Wisconsin and an assistant professor of adolescent medicine at the Medical College of Wisconsin.

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Pediatric Dermatology
Keisha Adams, MD

Pediatric Dermatology
Valerie Carlberg, MD

Pediatric Dermatology
Leah Lalor, MD

NEW ON STAFF
Specialists in our network ready to help
To refer a patient, call (800) 266-0366.
Ravit Boger, MD, is medical director of pediatric infectious disease at Children’s Hospital of Wisconsin and chief professor of pediatric infectious diseases at the Medical College of Wisconsin. 

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Samuel Adams, MD, is a pediatric neurologist at Children’s Hospital of Wisconsin and an assistant professor of pediatric neurology at the Medical College of Wisconsin.

KEY TO SYMBOLS: DEGREE RESIDENCY FELLOWSHIP BOARD CERTIFICATION
NEW ON STAFF

**Ophthalmology**

Smith Ann Chisholm, MD, is a pediatric ophthalmologist at Children’s Hospital of Wisconsin and an assistant professor in pediatric ophthalmology and visual sciences at the Medical College of Wisconsin. 🌐 Washington University in St. Louis School of Medicine, MD 🏥 Barnes Jewish Hospital/Washington University, Ophthalmology 🌐 University of Michigan Medical School, Pediatric Ophthalmology; Medical College of Wisconsin, Oculoplastics 💼 Ophthalmology

**Otolaryngology**

Sophie Shay, MD, is a pediatric otolaryngologist at Children’s Hospital of Wisconsin and an assistant professor of pediatric otolaryngology at the Medical College of Wisconsin. 🌐 University of Chicago, MD 🌐 UCLA Medical Center, Otolaryngology 🌐 Ann & Robert H. Lurie Children’s Hospital of Chicago, Pediatric Otolaryngology 💼 Pediatric Otolaryngology

**Psychiatry**

Erica Arrington, MD, is a pediatric psychiatrist at Children’s Hospital of Wisconsin and an assistant professor of child and adolescent psychiatry at the Medical College of Wisconsin. 🌐 University of North Carolina at Chapel Hill School of Medicine, MD 🌐 University of North Carolina Hospitals, Psychiatry 🌐 University of North Carolina Hospitals, Child and Adolescent Psychiatry 💼 Child and Adolescent Psychiatry

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**KEY TO SYMBOLS:** 🌐 DEGREE 🏥 RESIDENCY 🎯 FELLOWSHIP 🎪 BOARD CERTIFICATION
Retirements

Children’s Hospital of Wisconsin thanks these providers for their years of service.

Patricia Burrows, MD, Imaging
Bruce Camitta, MD, Hematology/Oncology
Steven Werlin, MD, Gastroenterology

Departures

Children’s Hospital of Wisconsin would like to thank the following providers for their contributions. We wish them well in future endeavors.

Omar Ali, MD, Endocrinology
Cheryl Cameron, MD, Emergency Medicine
Tina Damarjian, MD, Ophthalmology
Ankur Datta, MD, Neonatology
Nada Derar, MD, Genetics
Robert Newby, PhD, Neuropsychology
Kathy Russeth, MD, Psychiatry
David Smith, MD, Special Needs
Richard Stewart Hill, MD, Hospital Medicine
Monica Thaker, MD, Hematology/Oncology

Psychology

Kristin Hoff, PsyD, is a pediatric psychologist in orthopedic surgery at Children’s Hospital of Wisconsin and an assistant professor of orthopedic surgery at the Medical College of Wisconsin.
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- Ohio State University
- Psychology

Surgery

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- Johns Hopkins University School of Medicine, General Surgery
- Vanderbilt University Children’s Hospital, Pediatric Surgery
- General Surgery, Pediatric Surgery

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- Urology

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- Urology

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Steven Werlin, MD, Gastroenterology
Kenosha Clinic offers care close to home

Children’s Hospital of Wisconsin’s new Kenosha Clinic opened in February. The newly constructed building offers a comfortable, accessible environment and quality pediatric care in these specialty areas:

- Adolescent Medicine
- Asthma Allergy
- Cardiology
- Dermatology
- ENT
- GI
- Neurology
- Primary Care
- PT/OT
- Rehab
- Speech
- Urgent Care
- Urology

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Call the Physician Referral and Consultation line at (800) 266-0366.

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Kenosha, WI

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Saturday and Sunday
9 a.m. to 9 p.m.

ULTRASOUND
Monday-Thursday
7:30 a.m. to 6 p.m.
Friday
8 a.m. to 4:30 p.m.

URGENT CARE
7 days a week
9 a.m. to 9 p.m.