



# NeonatalNews.Net<sup>sm</sup>

From the Section of Neonatology, Department of Pediatrics, Baylor College of Medicine, Houston, Texas

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## Spotlight



### Doctor Karen Johnson

is an Assistant Professor of Pediatrics at Baylor College of Medicine, working as a faculty member in the Section of Neonatology serving hospitals throughout the Texas Medical Center. She received her Bachelor of Science Degree from Xavier University of Louisiana in 1978. She then attended Baylor College of Medicine earning a Doctor of Medicine in 1981. She continued her postdoctoral training at Baylor, completing her residency training in 1984 and fellowship training in 1986.

Currently, Dr. Johnson is a student at the University of Texas Health Science Center pursuing a Masters Degree in Public Health. She is Board certified in Pediatrics and Neonatal-Perinatal Medicine.

Dr. Johnson's community involvement includes volunteering at the Houston's Taping for the Blind, Sheltering Arms Senior Services, and the Gulf Coast March of Dimes. She is a past member of the Southwest YMCA Board of Directors. Dr. Johnson also is a familiar face on Houston's KHOU Channel 11, where she is the Medicine Correspondent for the "Focus on Health" television program.

Dr. Johnson is an outstanding clinician, educator and humanitarian who enjoys the challenges of academic medicine and any efforts to improve the health and well-being of children and of the community as a whole.

## The Front Line *Congenital heart surgery: New challenges for an evolving specialty*

by Charles D. Fraser, Jr., MD  
Professor of Congenital Heart Surgery



Charles D. Fraser, Jr., MD

Dramatic advances in technology and outcomes were seen in the field of congenital cardiac surgery over the last decade. Conditions with previously dismal prognoses are now handled with great success in large-volume centers committed to the care of patients with congenital heart anomalies. At Texas Children's Hospital, a combination of extensive experience, institutional commitment, and dedicated personnel affords our patients the opportunity of optimum care and every possibility of a quality outcome.

Expectations for children born with complex cardiac malformations continue to improve. Unfortunately, tremendous interinstitutional differences in survival rates remain. The general medical community does not largely recognize this fact and parents can face difficulty obtaining accurate information about an institution's performance. Peer reviewed medical publications remain the gold standard by which centers should be measured, as well as state-specific outcomes data such as are available in Texas. Parents and practitioners making decisions about patient referrals should be familiar with current performance information.

Many of the straightforward conditions, including patent ductus arteriosus and atrial septal defect, frequently are handled now through catheter-based, interventional approaches. Although they are surgically challenging, even small, premature neonates with complex cardiac malformations often are best treated with prompt, aggressive repair early in life. While these patients have additional physiologic derangements, surgery may be mandatory to avoid an unsalvageable situation. At the other extreme, it is becoming increasingly clear that adults with congenital heart disease requiring surgery often should be treated at a children's hospital.

Prenatal diagnosis and maternal referral allows early consultation with the surgical, pediatric cardiology, and neonatology teams at Texas Children's Hospital. This team approach greatly benefits the patient, in many cases avoiding serious perinatal complications. This, in turn, translates into a preoperative patient with the optimum physiologic state at the time of surgery.

At Texas Children's, surgical survival rates now approach 100% for many previously fatal cardiac malformations, including transposition of the great arteries, interrupted

## In Memoriam



**Murdina M. Desmond, MD** (1916–2003)  
Head, Section of Neonatology,  
Baylor College of Medicine  
1957–1972

Our faculty and staff were saddened to learn that the beloved Dr. Murdina Desmond had passed away in July. Her contributions both to the Section of Neonatology at Baylor College of Medicine and to the science and knowledge of the field of newborn medicine were remarkable.

“Dr. Murdina Desmond was a renowned and respected leader in neonatology worldwide,” said Dr. Leonard E. Weisman, current Head of Neonatology at Baylor. “Her research accomplishments were extensive, but none more than her work on the newborn’s transition from fetal to neonatal life. This work was a major milestone in neonatology and continues to enhance the care of all babies today.

“Murdina was an outstanding medical educator and everyone enjoyed spending time with her because of her friendly and outgoing personality. She gave birth to Neonatology at Baylor College of Medicine, blazed the trail for those who followed, and was an inspiration to everyone who knew her. She was a brilliant scientist, a thought-provoking teacher, a wonderful leader, and a fabulous friend. We all are going to miss her, but were blessed to have known her.”

Dr. Desmond was featured in the Spotlight column of the inaugural issue of NeonatalNews.Net in July 2000, which is archived online at

[www.neonatalnews.net](http://www.neonatalnews.net).

## Research Highlights

### Grants/Funding

**Diane Anderson, PhD**, Leadership Excellence in Maternal Child Health Nutrition; Health Resources and Services Administration (HRSA)-Maternal and Child Health, \$632,500.

**Bhagavatula Moorthy, PhD**, Molecular Mechanisms of Cytochrome P4501A1 Expression; National Institutes of Health, \$210,694.

### Events

**James M. Adams, Jr., MD**, invited speaker at the University of Texas School of Nursing in August. Topic: *Management of Bronchopulmonary Dysplasia*.

**Gerardo Cabrera-Meza, MD**, invited participant in Peer Review Group Meeting to analyze a strategy for neonatal health in New Delhi, India in July. The group was established to review the strategy and to improve neonatal health in Southeast Asia, the population of which represents 25% of the global neonatal population.

**Chantal Lau., PhD**, invited lecturer at St. Vincent Hospital, Santa Fe, New Mexico in August. Topic: *Oral Feeding in Preterm Infants*.

**Michael E. Speer, MD**, presentation at meeting of the Advisory Committee to the Secretary of the Department of Health and Human Services in Washington, DC, in August. Topic: *Neonatal Encephalopathy*.

## Fellowship Program reaccredited

Another five years of continued accreditation was granted to the Neonatal-Perinatal Medicine Fellowship Program at Baylor College of Medicine from the Accreditation Council for Graduate Medical Education. Following a rigorous review process, maximum allowable accreditation was granted ensuring certification of the fellowship program until 2008. “To maintain this level of certification requires the support and contribution of each and every faculty, fellow, and staff member and attests to the quality of our program,” said Dr. Leonard E. Weisman, the program director and Head of the Section of Neonatology at Baylor.

In 1961, Dr. Arnold J. (Jack) Rudolph began the fellowship program, and it was one of the first programs to be accredited by the ACGME in 1984. Ours is one of 98 Neonatal-Perinatal Medicine programs in the U.S. and has as many as 15 full-time fellows in training at any one time. The 150 fellows who have graduated the program account for over four percent of the neonatologists in the nation. During the last five years, fellows in training have published more than 70 peer-reviewed articles, presented their research at hundreds of national and regional scientific meetings, and received numerous awards and national recognition including Young Investigator Awards from the American Academy of Pediatrics, Society for Pediatric Research, Oxygen Society, and others.

For information about the Baylor Neonatal-Perinatal Medicine Fellowship Program, visit our Web site ([www.neonate.net/Trainees/trainees.htm](http://www.neonate.net/Trainees/trainees.htm)) or see the Fellowship box on page 4 of this newsletter.

## The Reba Michels Hill Lectureship

featuring

**Joseph Garcia-Prats, MD, & Cathy Garcia-Prats**

*The Joys and Challenges of Parenting*

November 6, 2003, 7:00 – 8:00 PM

Children’s Museum of Houston (Brown Auditorium) • 1500 Binz St. • Houston

RSVP to 713.522.1138 ext. 258 or [csprueill@cmhouston.org](mailto:csprueill@cmhouston.org)

AND

November 7, 2003, 8:30 – 9:30 AM

Pediatrics Grand Rounds

Texas Children’s Hospital (Auditorium) • 6621 Fannin St. • Houston

Presented by the Neonatology Service at Texas Children’s Hospital  
Hosted by Baylor College of Medicine, Texas Children’s Hospital, and Children’s Museum of Houston



Heidi E. Karpen, MD

## Case Study

### *Ebstein syndrome*

by Heidi E. Karpen, MD  
Assistant Professor of Pediatrics

This 3023 gm infant was born at 38 weeks' gestation by repeat cesarean section to a 22-year-old gravida III para II mother. Initially, the baby was admitted to the newborn nursery but was noted to be cyanotic at six hours of life. Physical exam revealed an infant in moderate respiratory distress with a 3/6 holosystolic murmur loudest at the RSB and a room air pulse oximetry of 69%. Arterial blood gas (ABG) on 100% oxyhood showed pH 7.34 pCO<sub>2</sub> 41 pO<sub>2</sub> 27. Chest roentgenogram (CXR) revealed a cardiac silhouette that occu-

piated >90% of the thoracic cavity. Patient was intubated and prostaglandin (PGE<sub>1</sub>) was started. Follow-up ABG on endotracheal CPAP and 100% oxygen was similar to that in room air.

On the second day of life (DOL), an echocardiogram revealed apical displacement of the septal leaflet of the tricuspid valve, which was severely limited by tethering to the interventricular septum. The anterior leaflet also was tethered but less so. These anomalies resulted in significant tricuspid regurgitation, producing severe right atrial dilation. The pulmonary valve was hypoplastic with minimal antegrade flow and trivial regurgitation. A large patent ductus arteriosus (PDA) was noted with left-to-right shunting. The right ventricle was small with severe hypertrophy but with qualitatively good ventricular systolic function.

On DOL 4, cardiac catheterization confirmed the echocardiographic findings with a pulmonary balloon valvuloplasty. Over the next several weeks, the infant was weaned from support and was discharged on oral furosemide. Discharge CXR showed marked reduction in the size of the cardiac silhouette.

### Discussion

Ebstein anomaly is a congenital cardiac defect consisting of three main components—severe malformation of the tricuspid valve, atrialization of the right ventricular wall, and patent foramen ovale. Ebstein anomaly may occur as an isolated defect or in conjunction with coarctation of the aorta or L-transposition of the great arteries. Wolff-Parkinson-White syndrome, or concealed bypass tracts, is seen in about 20% of patients.

Neonates with severe Ebstein anomaly typically present with cyanosis and signs of right heart failure. The murmur usually is a low-frequency holosystolic murmur of tricuspid regurgitation. ECG typically shows right bundle branch block with or without right atrial enlargement. Chest radiograph shows marked cardiomegally due to right atrial enlargement. The extent of cardiac enlargement is a useful tool in predicting severity and outcome. In mild forms, the diagnosis may be suspected during evaluation of a click or SVT.

The factors most strongly associated with poor outcome are diagnosis in the fetal or neonatal period, echocardiographic evidence of right ventricular outflow tract obstruction, and cardiothoracic ratio on CXR >90%.

Mild forms of Ebstein anomaly do not require any specific therapy and generally do well. Severely affected infants may have little prograde pulmonary flow and hence are ductal dependent. For these patients, therapy may require prolonged prostaglandin therapy or ECMO with subsequent surgical intervention. Treatment with iNO and balloon valvuloplasty offers a minimally invasive alternative and may delay the need for more aggressive intervention.

### References

1. van Son JA, Konstantinov IE, Zimmermann V, Wilhelm Ebstein and Ebstein's malformation. *Eur J Cardiothorac Surg* 2001 Nov;20(5):1082-1085.
2. Celermajer DS, Bull C, Till JA, Cullen S, et al. Ebstein's anomaly: presentation and outcome from fetus to adult. *J Am Coll Cardiol* 1994 Jan;23(1):170-176.

## Contact Us

The Baylor College of Medicine Section of Neonatology has staff at hospitals in Houston's Texas Medical Center and in the local community.

*To request a neonatal consultation at any of our locations, call*

**1-877-NEONATE**  
**(1-877-636-6283)**

### Texas Medical Center locations

#### **Texas Children's Hospital**

6221 Fannin Street, Houston TX 77030  
Director of Nurseries: James M. Adams, MD

**For neonatal transport, call the Kangaroo Crew:**

In Houston: 832-824-5550  
Toll-free: 1-877-770-5550

#### **St. Luke's Episcopal Hospital**

6720 Bertner Avenue, Houston TX 77030  
Director of Nurseries: Michael E. Speer, MD

#### **The Methodist Hospital**

6565 Fannin Street, Houston TX 77030  
Director of Nurseries: Michael E. Speer, MD

#### **Ben Taub General Hospital**

1504 Taub Loop, Houston TX 77030  
Director of Nurseries: Joe Garcia-Prats, MD

### Community locations

#### **East Houston Regional Medical Center**

13111 East Freeway, Houston TX 77015  
Director of Nurseries: Dilicia A. McLenan, MD

#### **St. Luke's Community Medical Center - The Woodlands**

17200 St. Luke's Way,  
The Woodlands TX 77384  
Director of Nurseries: Charles T. Hankins, MD

#### **Twelve Oaks Medical Center - Sharpstown**

6700 Bellaire Blvd, Houston TX 77074  
Director of Nurseries: Tommy Leonard, MD

#### **Woman's Hospital of Texas**

7600 Fannin Street, Houston TX 77056

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## Surgery (continued from page 1)

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aortic arch, total anomalous pulmonary venous return, and truncus arteriosus. Other lesions, historically associated with nearly uniform mortality, are now treated with ever improving success rates. Hypoplastic left heart syndrome (HLHS) remains the most common condition in newborns with congenital heart disease involving a single ventricle. Using a multidisciplinary approach, the operative mortality for surgical palliation of this difficult and common problem has steadily declined at Texas Children's, where the operative survival for children undergoing first stage surgical palliation for HLHS during the last three years has been greater than 90%.

For a small subset of patients, no therapy exists except cardiac transplantation. The world's first pediatric cardiac transplant procedure was done at Texas Children's, and we have a very large body of experience with this therapy.

Neurodevelopment is an area of intense research in children with congenital heart disease. Patients with cyanotic congenital heart disease palliated with the Fontan operation have been noted historically to have elevated risk of delayed development and decreased ultimate neurocognitive function. The precise etiology of this remains uncertain. The need for heart-lung bypass has been the suspected source of neurologic dysfunction in some. Using novel approaches, including a dedicated pediatric cardiac perfusion team, we monitor cerebral blood flow and brain oxygenation during bypass to precisely regulate perfusion during the entire operation.

Dedicated congenital heart centers, such as ours, will continue to lead the way in advances in therapy and patient outcomes.

### Baylor College of Medicine Neonatal Fellowships

#### Neonatal-Perinatal Medicine Fellowship Program

accepts applications year-round

**For information**

- visit our Web site: [www.neonate.net](http://www.neonate.net)
- send E-mail to:  
[fellowship-program@neo.bcm.tmc.edu](mailto:fellowship-program@neo.bcm.tmc.edu)

#### Neonatal Nutrition Fellowship Program

for Registered Dietitians with clinical experience;  
accepts applications year-round for two training  
periods (January-March and April-June).

**For information**

- visit our Web site: [www.neonate.net](http://www.neonate.net)

Or contact Diane Anderson, PhD, RD

- E-mail: [diane@bcm.tmc.edu](mailto:diane@bcm.tmc.edu)
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