

## Division Data Summary

### Research and Training Details

|                                     |           |
|-------------------------------------|-----------|
| Number of Faculty                   | 12        |
| Number of Joint Appointment Faculty | 4         |
| Number of Research Fellows          | 2         |
| Number of Support Personnel         | 55        |
| Direct Annual Grant Support         | \$274,951 |
| Peer Reviewed Publications          | 26        |

### Clinical Activities and Training

|                            |        |
|----------------------------|--------|
| Number of Clinical Staff   | 1      |
| Number of Clinical Fellows | 6      |
| Number of Other Students   | 4      |
| Inpatient Encounters       | 896    |
| Outpatient Encounters      | 47,019 |

## Division Photo



Row 1: S Shott, C Myer III, R Cotton, R Elluru, D Brown, M Rutter  
Row 2: E Arjmand, A deAlarcon, JP Willging, J Greinwald Jr, D Choo, K Johnson

## Significant Publications

Ahmed ZM, Yousaf R, et al. **Functional null mutations of MSRB3 encoding methionine sulfoxide reductase are associated with human deafness DFNB74.** *Am J Hum Genet.* 88(1): 19-29. 2011.

This paper describes the identification of first Methionine Sulfoxide Reductase (MSRB3) mutations causing isolated hearing loss in humans. Although MSRB3 is ubiquitously expressed in human physiology, hearing loss is the only symptom in families segregating MSRB3 mutations. This ostensible incongruity may reflect the uniquely high metabolic demand in the ear. This identification provides new insights about the function and the role of oxidative stress on the physiology of inner ear.

## Division Highlights

### Saima Riazuddin, PhD

Dr. Riazuddin was given the COMSTECH Ibrahim Memorial Award from the Islamic Academy of Sciences for her outstanding contributions to the field of research.

### Alessandro deAlarcon, MD and Ravi Elluru, MD, PhD

Dr. Jeffrey Houlton, Dr. Alessandro deAlarcon and Dr. Ravi Elluru submitted their work on Voice Outcomes Following Adult Cricotracheal Resection at the spring meetings. That work was awarded the Resident Research Award by the American Laryngological Association.

## Faculty Members

**Robin T. Cotton, MD**, Professor

*Director, Department of Pediatric Otolaryngology; Director, Aerodigestive and Sleep Center*

**Research Interests**

**Ellis M. Arjmand, MD, PhD**, Associate Professor  
*Director, Ear and Hearing Center*

**Research Interests**

**David K. Brown, PhD**, Assistant Professor  
*Director, Audiological Research Lab*

**Research Interests**

**Daniel I. Choo, MD**, Associate Professor  
**Research Interests**

**Alessandro deAlarcon, MD**, Assistant Professor  
*Director, Voice Clinic*

**Research Interests**

**Ravindhra G. Elluru, MD, PhD**, Assistant Professor  
**Research Interests**

**John. H. Greinwald Jr., MD**, Associate Professor  
**Research Interests**

**Charles M. Myer III, MD**, Professor  
*Director, Pediatric Otolaryngology Residency Program*

**Research Interests**

**Saima Riazuddin, PhD**, Assistant Professor  
*Director, Laboratory of Molecular Genetics*

**Research Interests**

**Michael J. Rutter, MD**, Associate Professor  
**Research Interests**

**Sally R. Shott, MD**, Professor  
**Research Interests**

**J. Paul Willging, MD**, Professor  
*Director, Pediatric Otolaryngology Fellowship Program*

**Research Interests**

## Joint Appointment Faculty Members

**Zubair Ahmed, PhD**, Assistant Professor  
Ophthalmology

**Research Interests** Genetics

**Jareen Meitzen-Derr, MPH, PhD**, Assistant Professor  
Biostatistics & Epidemiology

**Research Interests** Epidemiology

**Scott Holland, PhD**, Professor  
Neuroimaging Research Consortium

**Research Interests** Neuroimaging

**Dimitar Deliyski, PhD**, Associate Professor

Communication Science Research Center

**Research Interests** Communication Disorders

## Clinical Staff Members

- Michael Bowen, PA-C, RN, MA, Adult Airway

## Trainees

- Nicholas Smith, MD, PGYVI, University of Alabama-Birmingham
- Derek Lam, MD, MPH, PGYVI, University of Washington
- Catherine Hart, MD, PGYVI, University of Cincinnati
- Jonathan Ida, MD, PGYVII, Case Western Reserve University
- Kaalan Johnson, MD, PGYVII, Loma Linda University
- Stacey Clark, MD, PGYVII, The University of Texas Health Science Center

## Significant Accomplishments

### Oto-Gen

This past year has seen the development of a next-generation platform to rapidly sequence genes involved in pediatric hearing loss. This platform became a reality in the fall of 2011. Developed through a collaboration of the Ear and Hearing Center and the Molecular Genetics Laboratory at Cincinnati Children's, the "Oto-gen" will be the first NexGen sequencing platform to target common hearing loss genes. This technology will allow rapid and cost effective screening of genes that will diagnose and assist in treatment of pediatric patients with hearing loss.

### Hereditary Deafness

The auditory genetics lab of Saima Riazuddin, PhD, made large strides toward unlocking the mystery of hereditary deafness. Her lab recently identified the *MSRB3* gene that is responsible for autosomal recessively inherited deafness (DFNB74) in eight Pakistani families. In addition, her lab recently discovered a new locus for recessively inherited deafness (DFNB86) in another family. These discoveries could lead to diagnostic screening tools and treatments.

### Obstructive Sleep Apnea in Down Syndrome

A multidisciplinary project that successfully competed for National Institutes of Health funding looks to create and validate anatomical and physiological computational models of children with Down syndrome who have obstructive sleep apnea and use those models as predictors of success for surgical intervention.

## Division Publications

1. Ahmed ZM, Yousaf R, Lee BC, Khan SN, Lee S, Lee K, Husnain T, Rehman AU, Bonneux S, Ansar M, Ahmad W, Leal SM, Gladyshev VN, Belyantseva IA, Van Camp G, Riazuddin S, Friedman TB. **Functional null mutations of *MSRB3* encoding methionine sulfoxide reductase are associated with human deafness DFNB74.** *Am J Hum Genet.* 2011; 88:19-29.
2. Ali S, Riazuddin SA, Shahzadi A, Nasir IA, Khan SN, Husnain T, Akram J, Sieving PA, Hejtmancik JF,

- Riazuddin S. **Mutations in the beta-subunit of rod phosphodiesterase identified in consanguineous Pakistani families with autosomal recessive retinitis pigmentosa.** *Mol Vis.* 2011; 17:1373-80.
3. Borck G, Ur Rehman A, Lee K, Pogoda HM, Kakar N, von Ameln S, Grillet N, Hildebrand MS, Ahmed ZM, Nurnberg G, Ansar M, Basit S, Javed Q, Morell RJ, Nasreen N, Shearer AE, Ahmad A, Kahrizi K, Shaikh RS, Ali RA, Khan SN, Goebel I, Meyer NC, Kimberling WJ, Webster JA, Stephan DA, Schiller MR, Bahlo M, Najmabadi H, Gillespie PG, Nurnberg P, Wollnik B, Riazuddin S, Smith RJ, Ahmad W, Muller U, Hammerschmidt M, Friedman TB, Leal SM, Ahmad J, Kubisch C. **Loss-of-function mutations of ILDR1 cause autosomal-recessive hearing impairment DFNB42.** *Am J Hum Genet.* 2011; 88:127-37.
  4. Brown DK, Cameron S, Martin JS, Watson C, Dillon H. **The North American Listening in Spatialized Noise-Sentences test (NA LiSN-S): normative data and test-retest reliability studies for adolescents and young adults.** *J Am Acad Audiol.* 2010; 21:629-41.
  5. Cortina S, McGraw K, Dealarcon A, Ahrens A, Rothenberg ME, Drotar D. **Psychological Functioning of Children and Adolescents With Eosinophil-Associated Gastrointestinal Disorders.** *Child Health Care.* 2010; 39:266-278.
  6. Hamajima Y, Komori M, Preciado DA, Choo DI, Moribe K, Murakami S, Ondrey FG, Lin J. **The role of inhibitor of DNA-binding (Id1) in hyperproliferation of keratinocytes: the pathological basis for middle ear cholesteatoma from chronic otitis media.** *Cell Prolif.* 2010; 43:457-63.
  7. Hertzano R, Puligilla C, Chan SL, Timothy C, Depireux DA, Ahmed Z, Wolf J, Eisenman DJ, Friedman TB, Riazuddin S, Kelley MW, Strome SE. **CD44 is a marker for the outer pillar cells in the early postnatal mouse inner ear.** *J Assoc Res Otolaryngol.* 2010; 11:407-18.
  8. Ida JB, Mansfield S, Meinzen-Derr JK, Choo DI. **Complications in pediatric osseointegrated implantation.** *Otolaryngol Head Neck Surg.* 2011; 144:586-91.
  9. Kelchner LN, Brehm SB, Weinrich B, Middendorf J, deAlarcon A, Levin L, Elluru R. **Perceptual evaluation of severe pediatric voice disorders: rater reliability using the consensus auditory perceptual evaluation of voice.** *J Voice.* 2010; 24:441-9.
  10. Ksiazek J, Prager JD, Sun GH, Wood RE, Arjmand EM. **Inhaled cidofovir as an adjuvant therapy for recurrent respiratory papillomatosis.** *Otolaryngol Head Neck Surg.* 2011; 144:639-41.
  11. Manning PB, Rutter MJ, Lisek A, Gupta R, Marino BS. **One slide fits all: the versatility of slide tracheoplasty with cardiopulmonary bypass support for airway reconstruction in children.** *J Thorac Cardiovasc Surg.* 2011; 141:155-61.
  12. Meinzen-Derr J, Wiley S, Choo DI. **Impact of early intervention on expressive and receptive language development among young children with permanent hearing loss.** *Am Ann Deaf.* 2011; 155:580-91.
  13. Meinzen-Derr J, Wiley S, Grether S, Choo DI. **Children with cochlear implants and developmental disabilities: a language skills study with developmentally matched hearing peers.** *Res Dev Disabil.* 2011; 32:757-67.
  14. Neidich MJ, Prager JD, Clark SL, Elluru RG. **Comprehensive airway management of neonatal head and neck teratomas.** *Otolaryngol Head Neck Surg.* 2011; 144:257-61.
  15. Prager JD, Hopkins BS, Propst EJ, Shott SR, Cotton RT. **Oropharyngeal stenosis: a complication of multilevel, single-stage upper airway surgery in children.** *Arch Otolaryngol Head Neck Surg.* 2010; 136:1111-5.
  16. Prager JD, Myer CM, 3rd, Pensak ML. **Improving the letter of recommendation.** *Otolaryngol Head Neck Surg.* 2010; 143:327-30.
  17. Prager JD, Myer CMt, Hayes KM, Myer CM, 3rd, Pensak ML. **Improving methods of resident selection.** *Laryngoscope.* 2010; 120:2391-8.
  18. Propst EJ, Prager JD, Adams JM, Arjmand EM, Willging JP, Samy RN. **A preliminary investigation of four-dimensional ultrasound for evaluation of middle ear ossicles: an in vitro study.** *Int J Pediatr*

- Otorhinolaryngol.* 2010; 74:1028-33.
19. Propst EJ, Prager JD, Meinzen-Derr J, Clark SL, Cotton RT, Rutter MJ. **Pediatric tracheal reconstruction using cadaveric homograft.** *Arch Otolaryngol Head Neck Surg.* 2011; 137:583-90.
  20. Riazuddin S, Ahmed ZM, Hegde RS, Khan SN, Nasir I, Shaukat U, Butman JA, Griffith AJ, Friedman TB, Choi BY. **Variable expressivity of FGF3 mutations associated with deafness and LAMM syndrome.** *BMC Med Genet.* 2011; 12:21.
  21. Riazuddin SA, Shahzadi A, Zeitz C, Ahmed ZM, Ayyagari R, Chavali VR, Ponferrada VG, Audo I, Michiels C, Lancelot ME, Nasir IA, Zafar AU, Khan SN, Husnain T, Jiao X, MacDonald IM, Riazuddin S, Sieving PA, Katsanis N, Hejtmancik JF. **A mutation in SLC24A1 implicated in autosomal-recessive congenital stationary night blindness.** *Am J Hum Genet.* 2010; 87:523-31.
  22. Statham MM, Myer CM, 3rd. **Complications of adenotonsillectomy.** *Curr Opin Otolaryngol Head Neck Surg.* 2010; 18:539-43.
  23. Statham MM, Willging JP. **Automated high-level disinfection of nonchanneled flexible endoscopes: duty cycles and endoscope repair.** *Laryngoscope.* 2010; 120:1946-9.
  24. Sun GH, Harnych BM, Dickson JM, Gonzalez del Rey JA, Myer CM, 3rd, Greinwald JH, Jr.. **Characteristics of children diagnosed as having coagulopathies following posttonsillectomy bleeding.** *Arch Otolaryngol Head Neck Surg.* 2011; 137:65-8.
  25. Sun GH, Samy RN, Tinkle BT, Cornelius RS, Brown DK. **Craniometaphyseal dysplasia-induced hearing loss.** *Otol Neurotol.* 2011; 32:e9-10.
  26. Wootten CT, Shott SR. **Evolving therapies to treat retroglossal and base-of-tongue obstruction in pediatric obstructive sleep apnea.** *Arch Otolaryngol Head Neck Surg.* 2010; 136:983-7.

## Grants, Contracts, and Industry Agreements

| Grant and Contract Awards   | Annual Direct / Project Period Direct |
|---|---------------------------------------|
| <b>CHOO, D</b>  |                                       |
| <b>A Preclinical Trial of Intratympanic Antivirals for CMV-Related Hearing Loss</b> |                                       |
| National Institutes of Health   |                                       |
| R01 DC 008651   | 01/01/07-12/31/11                     |
|   | \$197,243                             |
| <b>Improved Method of Drug Delivery to the Inner Ear</b>                            |                                       |
| National Institutes of Health(University of Cincinnati)                             |                                       |
| R21 DC 011062   | 08/02/10-05/31/11                     |
|   | \$34,981                              |
| <b>RIAZUDDIN, S</b>   |                                       |
| <b>Defining the Role of Tricellular Tight Junction Protein</b>                      |                                       |
| Deafness Research Foundation  |                                       |
|   | 07/01/10-06/30/11                     |
|   | \$22,727                              |
| <b>Genetic and Functional Characterization of DFNB74 Gene</b>                       |                                       |
| The National Organization for Hearing Research Foundation                           |                                       |
|   | 01/28/11-01/27/12                     |
|   | \$20,000                              |
|   | <b>Current Year Direct</b>            |
|   | <b>\$274,951</b>                      |
|   | <b>Total</b>                          |
|   | <b>\$274,951</b>                      |