AudGenDB: a Public, Internet-Based, Audiologic - Otologic - Genetic Database for Pediatric Hearing Research

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AudGenDB Webinar

- Overview of AudGenDB
- Live Demonstration
  - General Capabilities
  - Specific case examples
- Questions from audience

Introduction

- Pediatric hearing research integrates complex data from varying specialties, on large numbers of patients.
- Examples of such data:
  - Audiogram
  - Evoked responses (e.g. ABR)
  - Speech and language performance
  - Medical and surgical history
  - Temporal bone imaging
  - Ancillary laboratory studies
  - Clinical genetics, dysmorphology
  - Genotype for known HL genes
  - Genomic data.
- Large-scale hearing research projects would benefit from an integrated electronic database that incorporates these varied data.

- Projects that could benefit from a large-scale database:
  - Risk factors for hearing loss progression
  - Study of rare patterns of hearing loss
  - Outcomes in mild or unilateral HL
  - Anatomy and genetics of inner ear malformations.
  - Identification of novel genes for distinct patterns of HL
  - Correlation of genotype with phenotype (audiologic data)

AudGenDB

At The Children's Hospital of Philadelphia (CHOP), we have developed just such a database –the AudGenDB.

Our ultimate goals are to:

- create a national repository of data for tens of thousands of children from a consortium of institutions
- make it freely available to researchers world-wide
- Protect the privacy of the included subjects.

Scope of AudGenDB

- Web-based, relational database
- Automatically extracts data from the electronic medical record - Audiologic, otologic, radiologic, genetic
- All patients who have undergone audiologic testing at CHOP since 2006.
  - 37,273 children
- Intuitive, web-based interface.
- Anonymized patient information.

Data Residing in AudGenDB

Audiology

- Complete audiograms, including speech and tympanometry (>57,000)
- ABR results (16,784 datasets)
- OAE data
- Use of hearing aids, cochlear implants

Otology

- Chronic medical diagnoses, otologic diagnoses
- Surgical procedures (>22,000)*

*upcoming release

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Data Residing in AudGenDB

Radiology

- Actual CT and MRI images (>1000 studies)
- Temporal bone CT and MRI reports (15,059 studies)*

Genetics

- Clinical genetic data – known genes (>1,000 patients)
- Existence of high-resolution genomic data (> 500 pts.)
- Access to raw genomic data, and links to original DNA samples.*

*upcoming release

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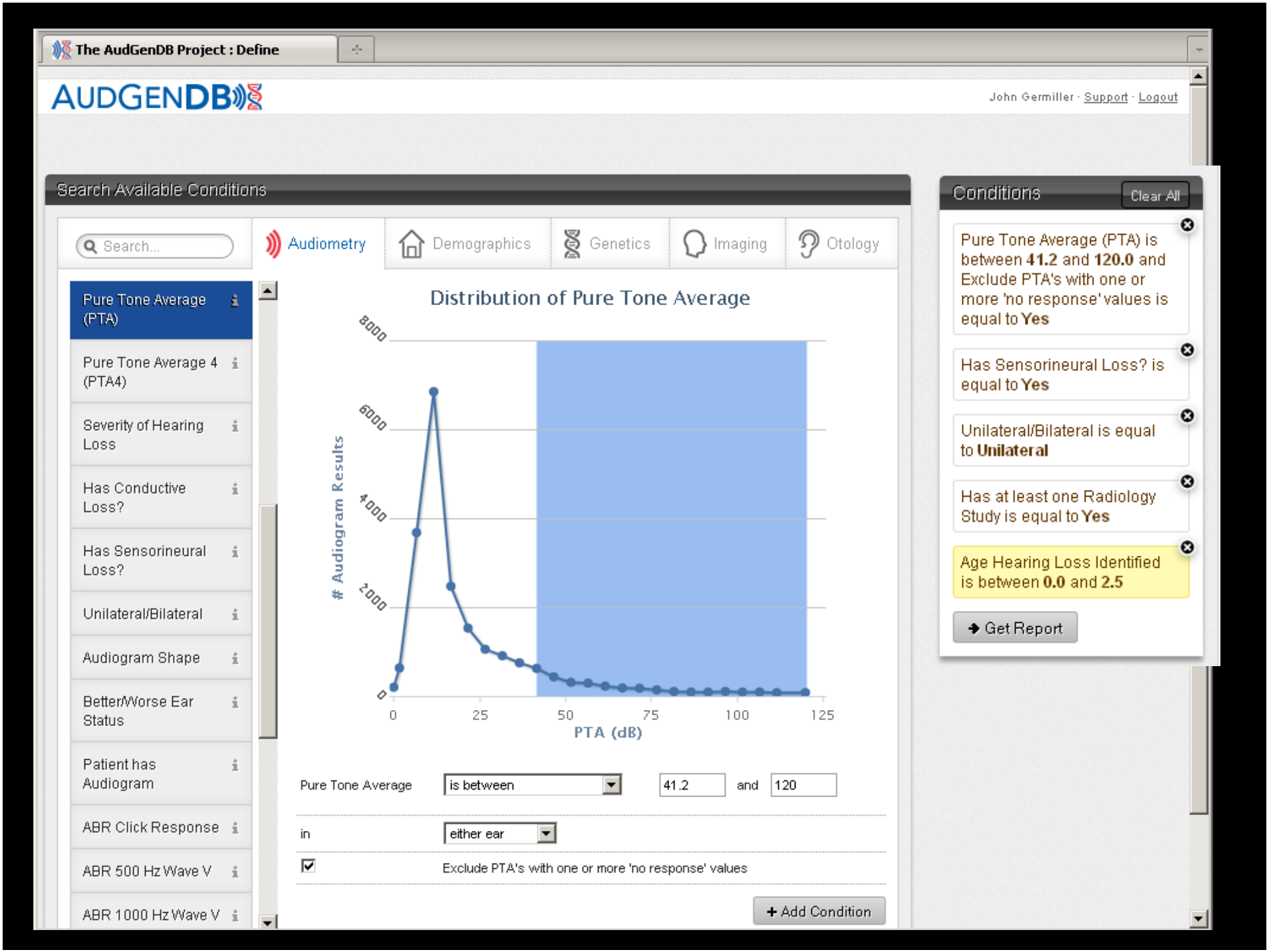
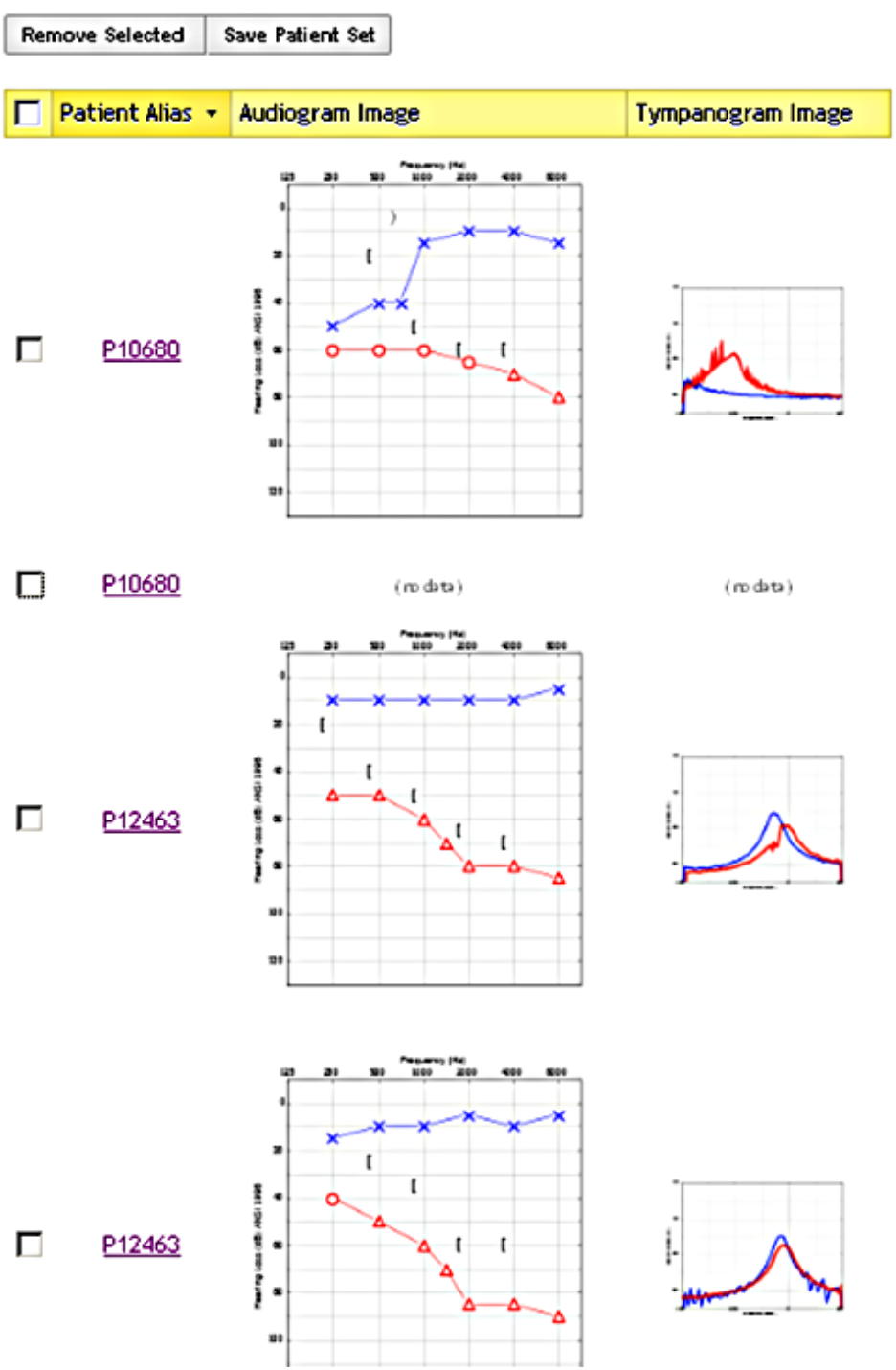
Searchable Criteria in AudGenDB

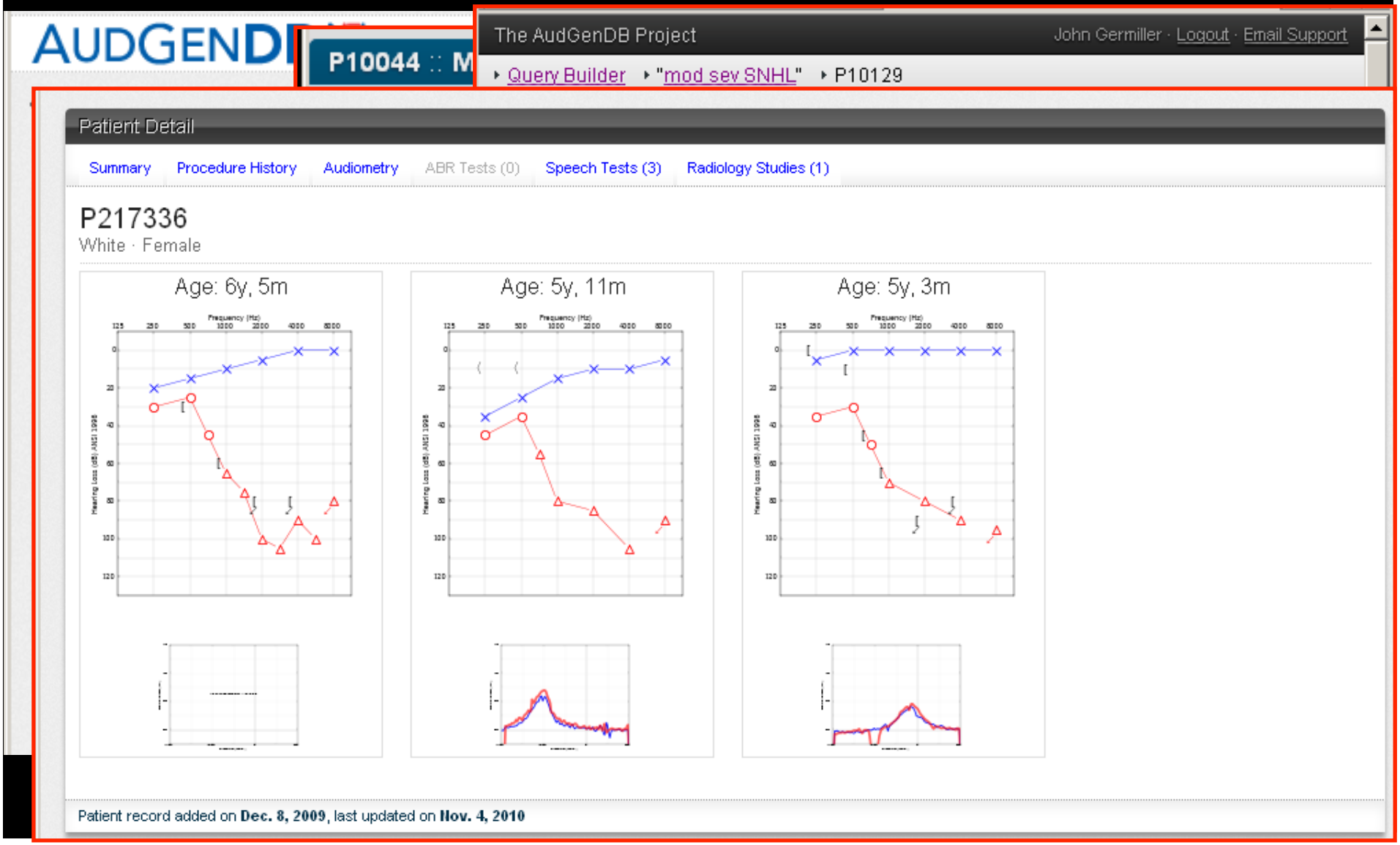
Category	Searchable Parameter	Example of Query
Demographic	Age, gender, ethnicity	Prelingual at diagnosis
Audiologic	Type of hearing loss	SNHL, conductive, mixed
	Laterality/ symmetry	Unilateral, Bilateral, Asymmetric
	Severity of loss	Mild hearing loss only Severe-profound only
	Loss at specific frequencies	High frequency HL
Otologic	Surgical	Identify patients who have undergone cochlear implantation; myringotomy tubes
	Medical	Bacterial meningitis
Radiologic	Existence of imaging	Identify only patients who have imaging studies available
Genetic	Genotype	SLC26A4 mutations GJB2 heterozygotes
	Existence of genomic SNP array data	Identify patients genotyped on whole-genome SNP arrays


The AudGenDB Project : Define
AUDGENDB
John Germiller · Support · Logout
Search Available Conditions
Search...
Audiometry
Demographics
Genetics
Imaging
Otology
Pure Tone Average (PTA)
Pure Tone Average 4 (PTA4)
Severity of Hearing Loss
Has Conductive Loss?
Has Sensorineural Loss?
Unilateral/Bilateral
Audiogram Shape
Better/Worse Ear Status
Patient has Audiogram
ABR Click Response
ABR 500 Hz Wave V
ABR 1000 Hz Wave V
Distribution of Pure Tone Average
Audiogram Results
PTA (dB)
0
2000
4000
6000
8000
0
25
50
75
100
125
Pure Tone Average
is between
41.2
and
120
in
either ear
Exclude PTA's with one or more 'no response' values
+ Add Condition
Conditions
Clear All
Pure Tone Average (PTA) is between 41.2 and 120.0 and Exclude PTA's with one or more 'no response' values is equal to Yes
Has Sensorineural Loss? is equal to Yes
Unilateral/Bilateral is equal to Unilateral
Has at least one Radiology Study is equal to Yes
Age Hearing Loss Identified is between 0.0 and 2.5
Get Report

Search Results Page



Patient-level data



Temporal Bone Imaging

- TB CT, TB MRI, IAC MRI, brain MRI, brain CT.
- Direct viewing of *original images*.
  - Modified DICOM viewer
  - scrolling through image stacks, adjust contrast, zoom.
  - length measurement (<0.1 mm).

- Radiologist reports - text files*

*upcoming release

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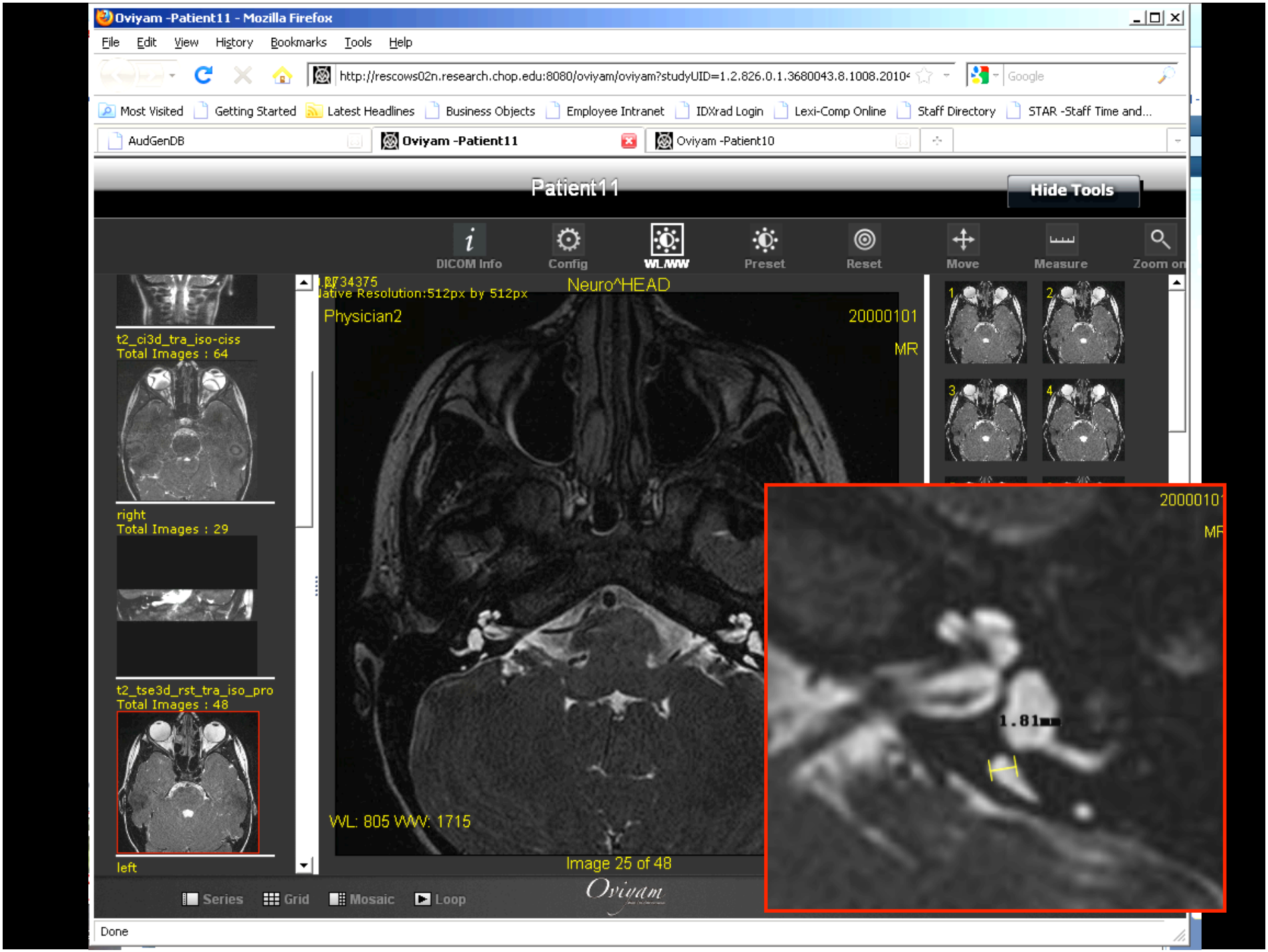
Oviyam -Patient11 - Mozilla Firefox
File Edit View History Bookmarks Tools Help
http://rescows02n.research.chop.edu:8080/oviyam/oviyam?studyUID=1.2.826.0.1.3680043.8.1008.2010
Most Visited Getting Started Latest Headlines Business Objects Employee Intranet IDXrad Login Lexi-Comp Online Staff Directory STAR -Staff Time and...
AudGenDB
Oviyam -Patient11
Oviyam -Patient10
Patient11
Hide Tools
DICOM Info
Config
WL/WW
Preset
Reset
Move
Measure
Zoom on
Native Resolution:512px by 512px
Neuro^HEAD
Physician2
20000101
MR
t2_ci3d_tra_iso-ciss
Total Images : 64
right
Total Images : 29
t2_tse3d_rst_tra_iso_pro
Total Images : 48
left
WL: 805 WW: 1715
Image 25 of 48
Series
Grid
Mosaic
Loop
Oviyam
1.81
Done

P10019 :: Male :: White

Summary | Procedure History | Audiometry | ABR Tests (0) | Speech Tests (3) | Radiology Studies (0) | Radiology Impressions (2)

⚠ The following data are direct excerpts from the individual radiologist's reports dictated at the time of the imaging. Radiology reports are subject to significant variability, due to variations in expertise and specialization, volume of neuroimaging from center to center, quality of the raw images, etc. Users are strongly encouraged to view and interpret the original images themselves, rather than relying solely on radiology reports. Systematic review by a pediatric neuroradiologist, with experience in inner ear imaging, is strongly recommended.

Age: 6y, 10m

CT OF THE TEMPORAL BONES [scrubbed date]

HISTORY: Sensorineural hearing loss.

COMPARISON: None.

TECHNIQUE: CT of the temporal bones acquired in the axial plane with 0.75 mm collimation images. Targeted small field of view reconstructions were obtained of each temporal bone, and coronal reformatted images.

FINDINGS: Bilateral myringotomy tubes are present. There is questionable finding of a minor abnormality involving both cochleae, with the apex appearing slightly full raising the possibility of subtle incomplete partitioning. It is unclear whether this finding is due to an actual anatomical abnormality, or perhaps due to the plane of scanning. The internal auditory canals are normal in size, as are the vestibular aqueducts bilaterally. The middle and external ear structures are normal, including the ossicular chains bilaterally.

The mastoid air cells are clear. There is incidental mucosal opacification in the left sphenoid sinus.

IMPRESSION:

No definite abnormality seen. However, there is a questionable abnormality involving the cochleae bilaterally with minimally incomplete partitioning at the apex. Correlation with audiometric testing is suggested.

70480, 76375 END OF IMPRESSION:

DD: [scrubbed date], [scrubbed time] DT: [scrubbed date], [scrubbed time]

Genetic Data

- Genotype for common HL genes – Connexin 26 and 30 (GJB2 ,GJB6), *Pendred (SLC26A4), *A1555G mitochondrial.
- Clinical SNP microarray results available in report form.
- Deletions or insertions (CNV) “calls” for >100 pts with SNHL*

*upcoming release

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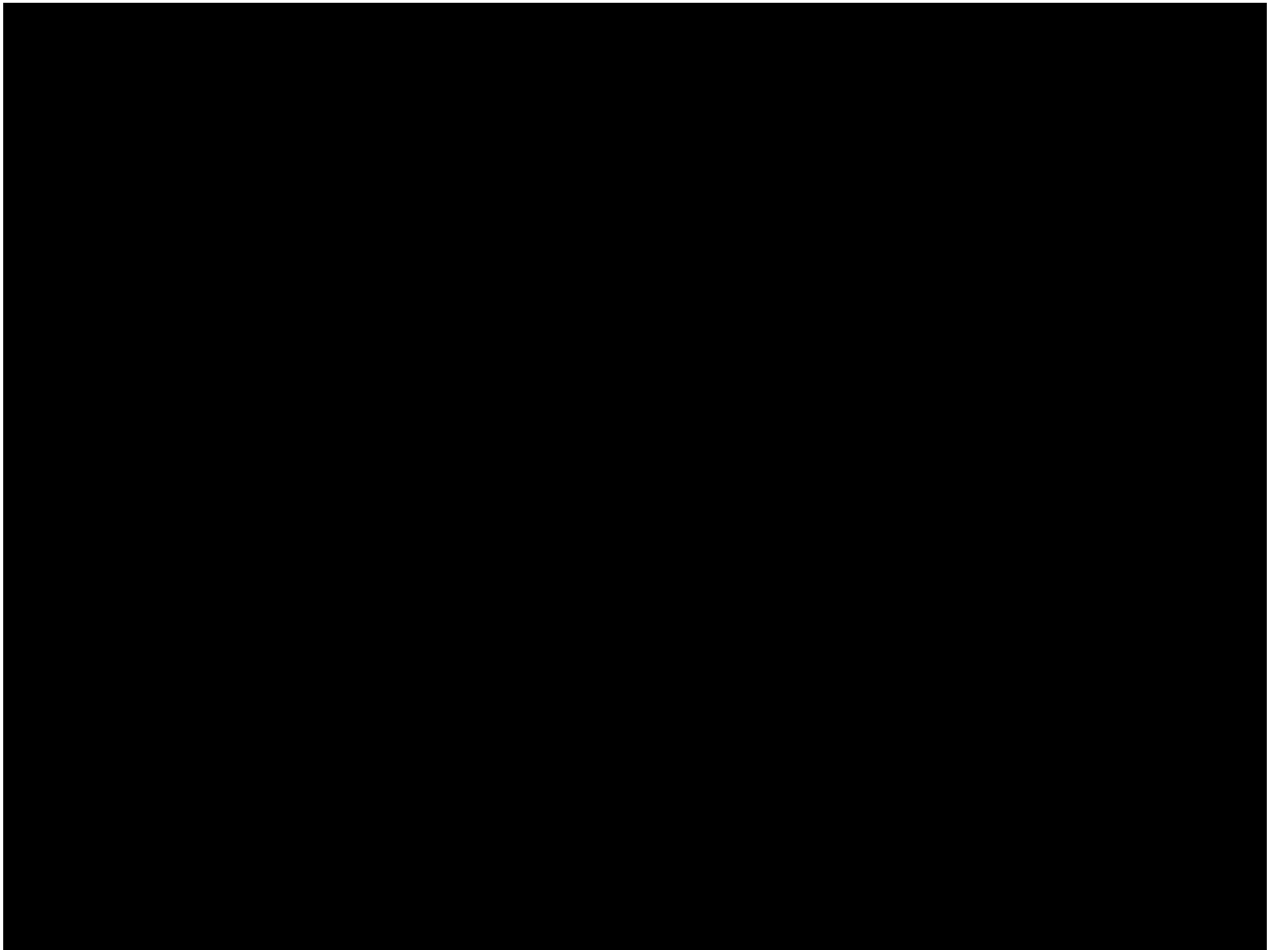
Genomic Data

- Goal: to provide access to raw genomic data (SNPs, next-generation sequencing)
- At this time, AudGenDB does yet not provide raw genomic data.
- However, flags to its existence allow the user to contact the CHOP investigator to request access to the data, or the original DNA.



A Public Data Resource

- Public release occurred in July 2011.
- Web-based. *Audgendb.chop.edu*
- Tutorials for new users.
- Access requires only a simple registration process, through the website.
- ***IRB approval is not necessary.***

(Access to original DNA would require IRB approval at both institutions)

The Future

- Plans for additional data
- Data from other institutions

Plans for Additional Data

Sooner:

- Raw ABR and OAE data
- Genomic Data
- EKGs (tracings and reports)
- Ancillary laboratory values

Later:

- UNHS data
- Risk factors for SNHL (e.g., prematurity, hyperbilirubinemia)
- Outcome data
- Temporal bone analysis

Data from Other Institutions

- A central goal has been to integrate data from other pediatric hospitals.
- Facilitated by our ongoing collaborations, Vanderbilt University will be first, followed by Children's Hospital Boston.

- *We seek other institutions who wish to make their data available for future incorporation into this resource.*

Conclusions

- AudGenDB represents the first large-scale database resource for pediatric hearing research.
- Contains detailed audiometric, otologic, radiologic, and genetic data - 37,000 children.
- AudGenDB is self-renewing and continually growing, as it automatically imports data monthly from the EMR.

Conclusions

- Intuitive web-based interface serves both beginner and advanced users.
- Available now to all researchers.
- AudGenDB to become multi-institutional with addition of Vanderbilt and Children's Hospital Boston.
- ***We welcome additional centers to help expand this resource nationally.***

  - ***John Germiller*** ***germiller@email.chop.edu***
  - ***Bryan Crenshaw*** ***crenshaw@email.chop.edu***

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Acknowledgments

- Supported by NIDCD Grant 5R33DC008630, *"Audiological and Genetic Resource for Pediatric Hearing Research,"* E. Bryan Crenshaw III, Principal Investigator.
- Initial phase supported by NIDCD 5R21DC008630, Judith S. Gravel, PhD, Principal Investigator.
- External Advisory Board
  - Audiology - Linda Hood PhD - Vanderbilt
  - Genetics and Otology - Margaret Kenna MD - Boston
  - Medical Informatics - John Pestian PhD – Cincinnati
- LaunchDM, LLC – website development
- Judy Gravel, Ph.D. (1948-2008), whose leadership at the outset of the project ensured its success.

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