Mass General Brigham
Mass Eye and Ear

Temporal Bone Image Library Chapter: Developmental Defects

Otopathology Laboratory

HARVARD MEDICAL SCHOOL
TEACHING HOSPITAL

Unaffected, L-191

Title: Unaffected, L-191

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA 9

TB Case Number: 231

Comments: Middle turn of cochlea

Gender: Male

Age (yrs.): 56

Otologic Diagnosis:

1. Temporal bone, normal, left
2. Membranous labyrinth, normal, left
3. Bony labyrinth, normal, left
4. Artifact, removal, amputation, cochlea, partial, left
5. Artifact, manipulative intentional infracture of footplate, left
6. Artifact, bone dust, saw-g

DFNA9, R-101

Title: DFNA9, R-101

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 398

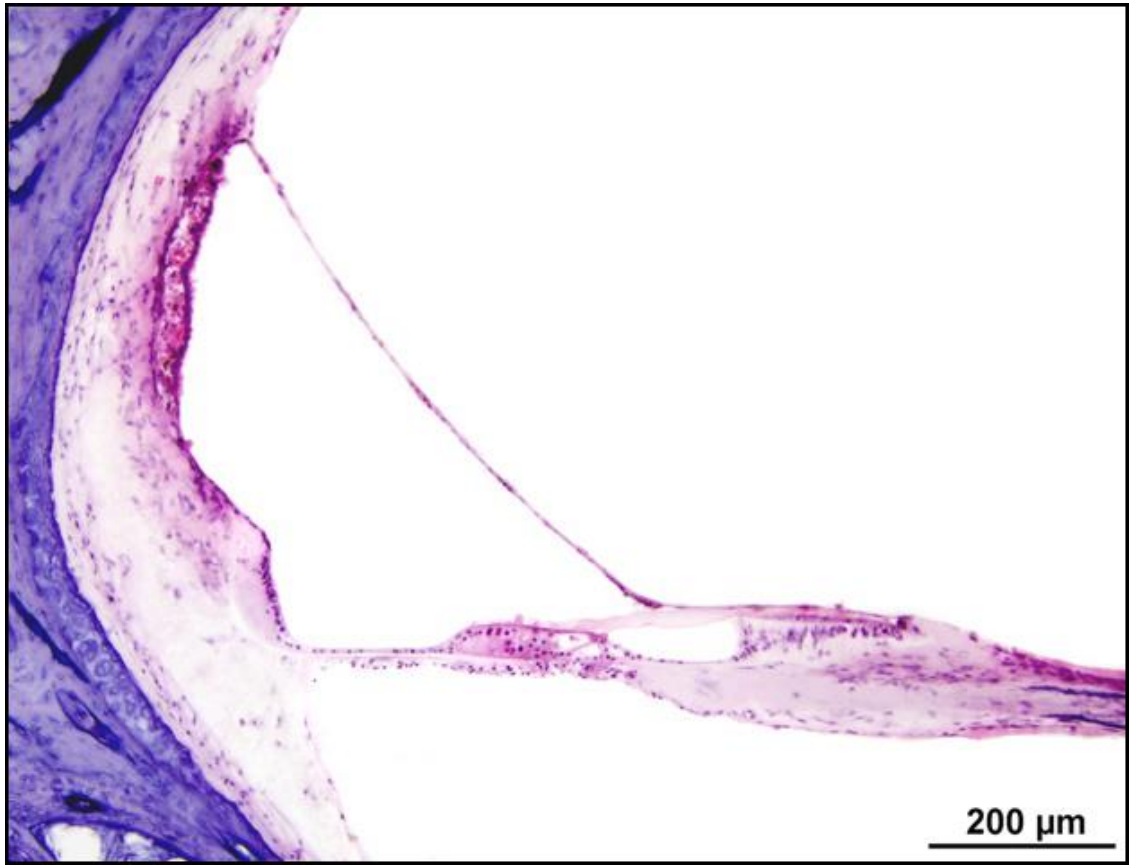
Comments: DFNA9 with COCH mutation, middle turn

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Atrophy, collapse and distortion of the vestibular membranous labyrinth, bilateral, probably caused by the effects of aging
2. Degeneration of the vestibular nerves, bilateral, associated with or secondary to:
    a) Presbycusis, neural type
    b) Tympanosclerosis, right ear



DFNA9, L-280

Title: DFNA9, L-280

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA 9

TB Case Number: 230

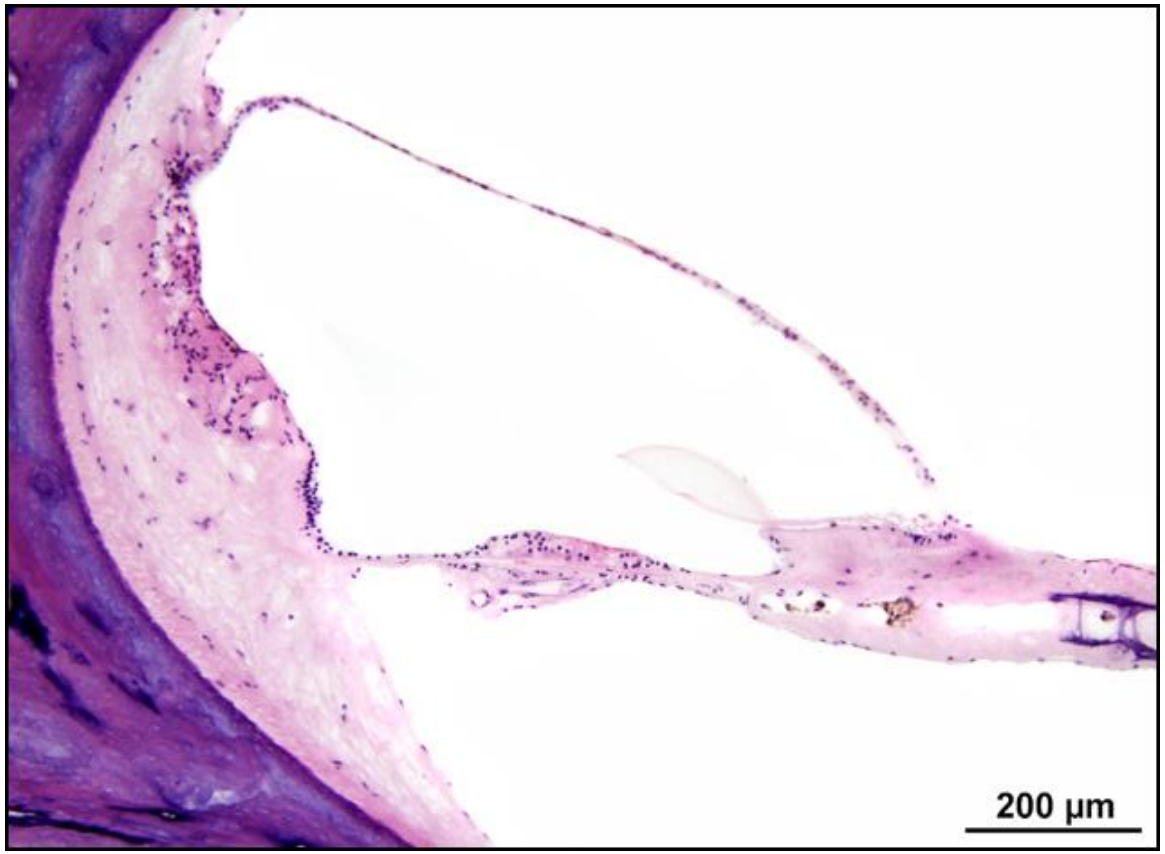
Comments: DFNA9 with COCH mutation, middle turn

Gender: Female

Age (yrs.): 86

Otologic Diagnosis:

1. Hereditary deafness, sensorineural, severe, adult-onset, autosomal dominant, bilateral
2. Deposit, ground substance, proliferation, auditory and vestibular sense organs, bilateral
3. Deposit, glycosaminoglycans, auditory and vestibular sense organs



Unaffected, L-191.2

Title: Unaffected, L-191.2

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 231

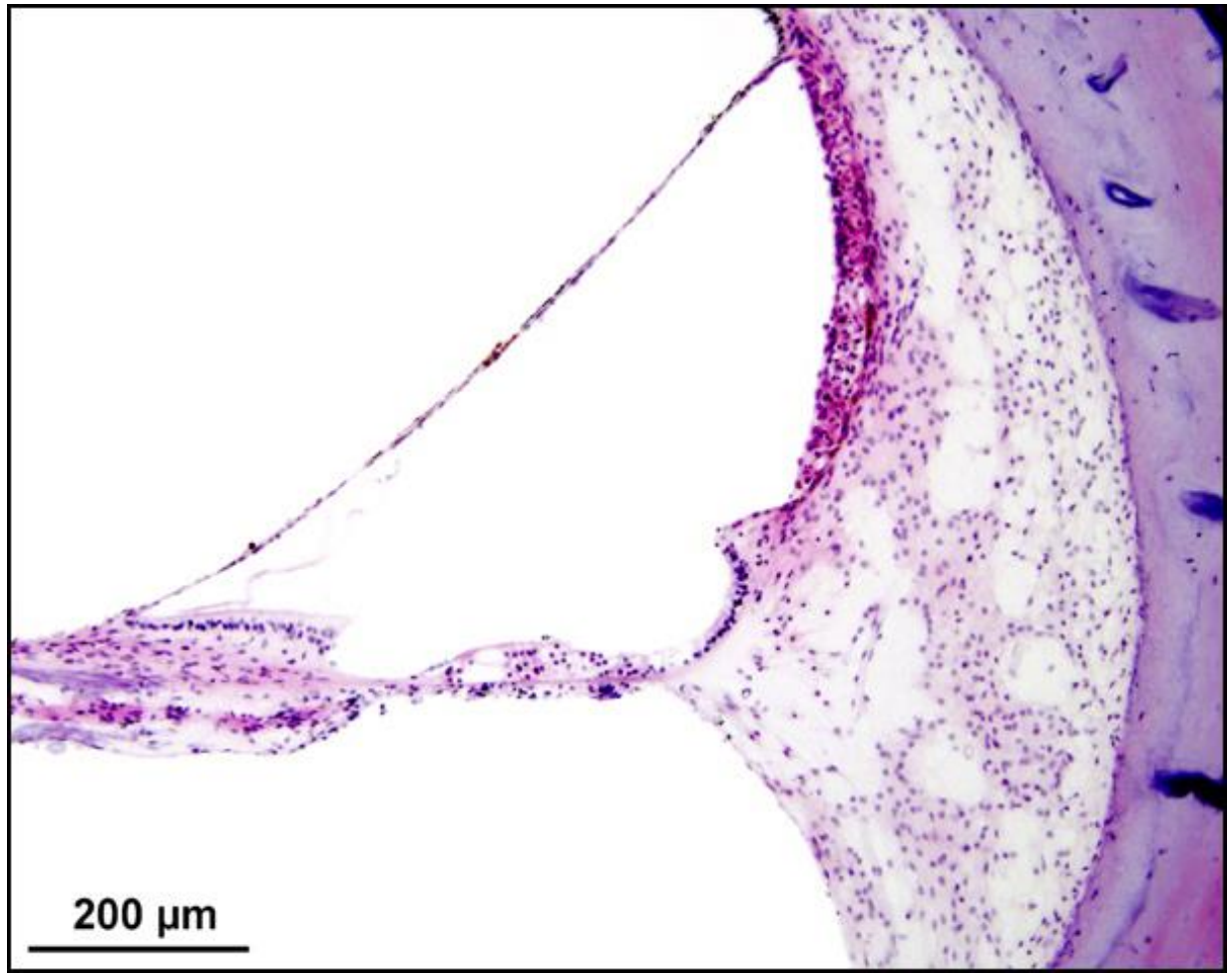
Comments: Basal turn

Gender: Male

Age (yrs.): 56

Otologic Diagnosis:

1. Temporal bone, normal, left
2. Membranous labyrinth, normal, left
3. Bony labyrinth, normal, left
4. Artifact, removal, amputation, cochlea, partial, left
5. Artifact, manipulative intentional infracture of footplate, left
6. Artifact, bone dust, saw-g



DFNA9, L-270

Title: DFNA9, L-270

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 230

Comments: DFNA9 with COCH mutation, basal turn

Gender: Female

Age (yrs.): 86

Otologic Diagnosis:

1. Hereditary deafness, sensorineural, severe, adult-onset, autosomal dominant, bilateral
2. Deposit, ground substance, proliferation, auditory and vestibular sense organs, bilateral
3. Deposit, glycosaminoglycans, auditory and vestibular sense organs

Unaffected, L-201

Title: Unaffected, L-201

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

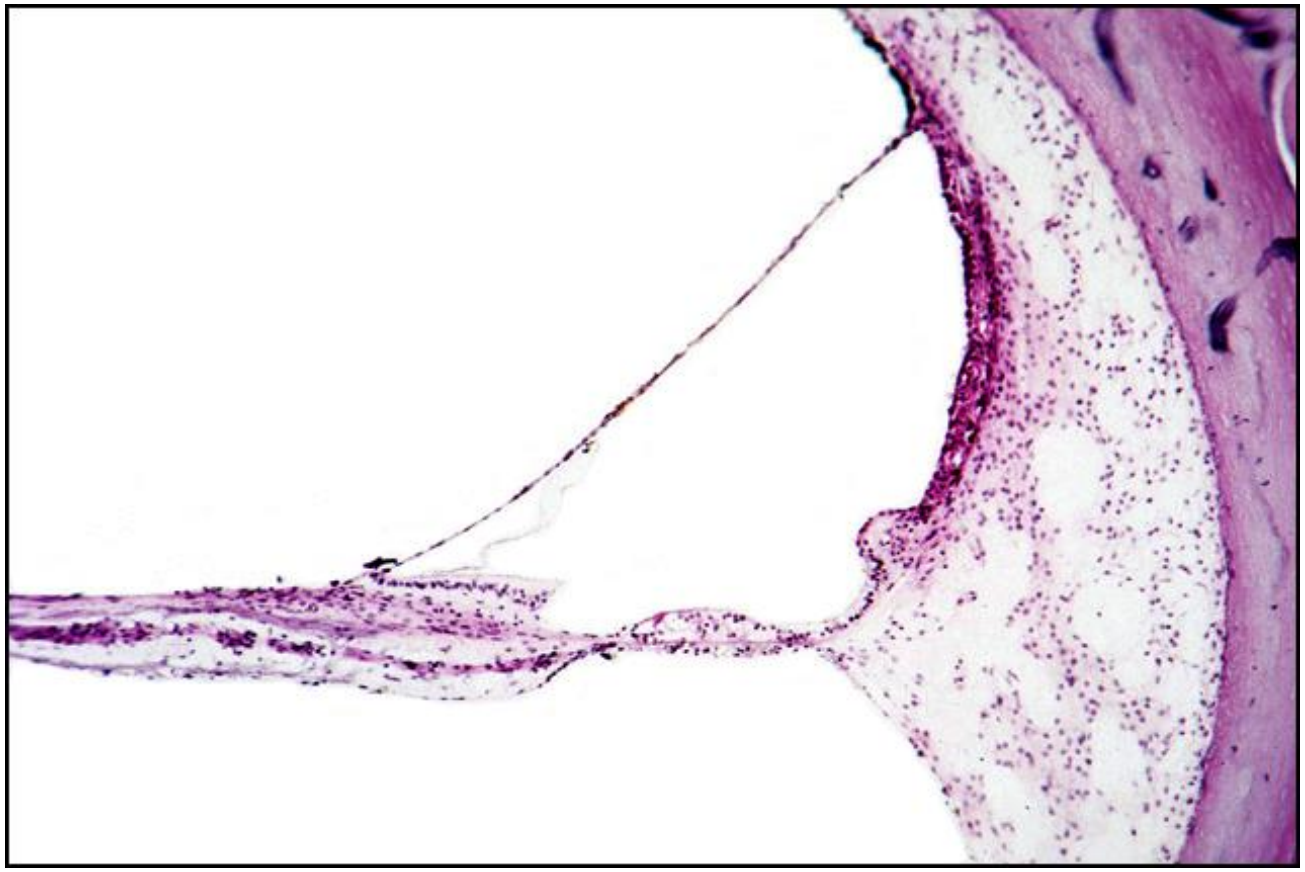
TB Case Number: 231

Gender: Male

Age (yrs.): 56

Otologic Diagnosis:

1. Temporal bone, normal, left
2. Membranous labyrinth, normal, left
3. Bony labyrinth, normal, left
4. Artifact, removal, amputation, cochlea, partial, left
5. Artifact, manipulative intentional infracture of footplate, left
6. Artifact, bone dust, saw-g



Unaffected, L-71

Title: Unaffected, L-71

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 231

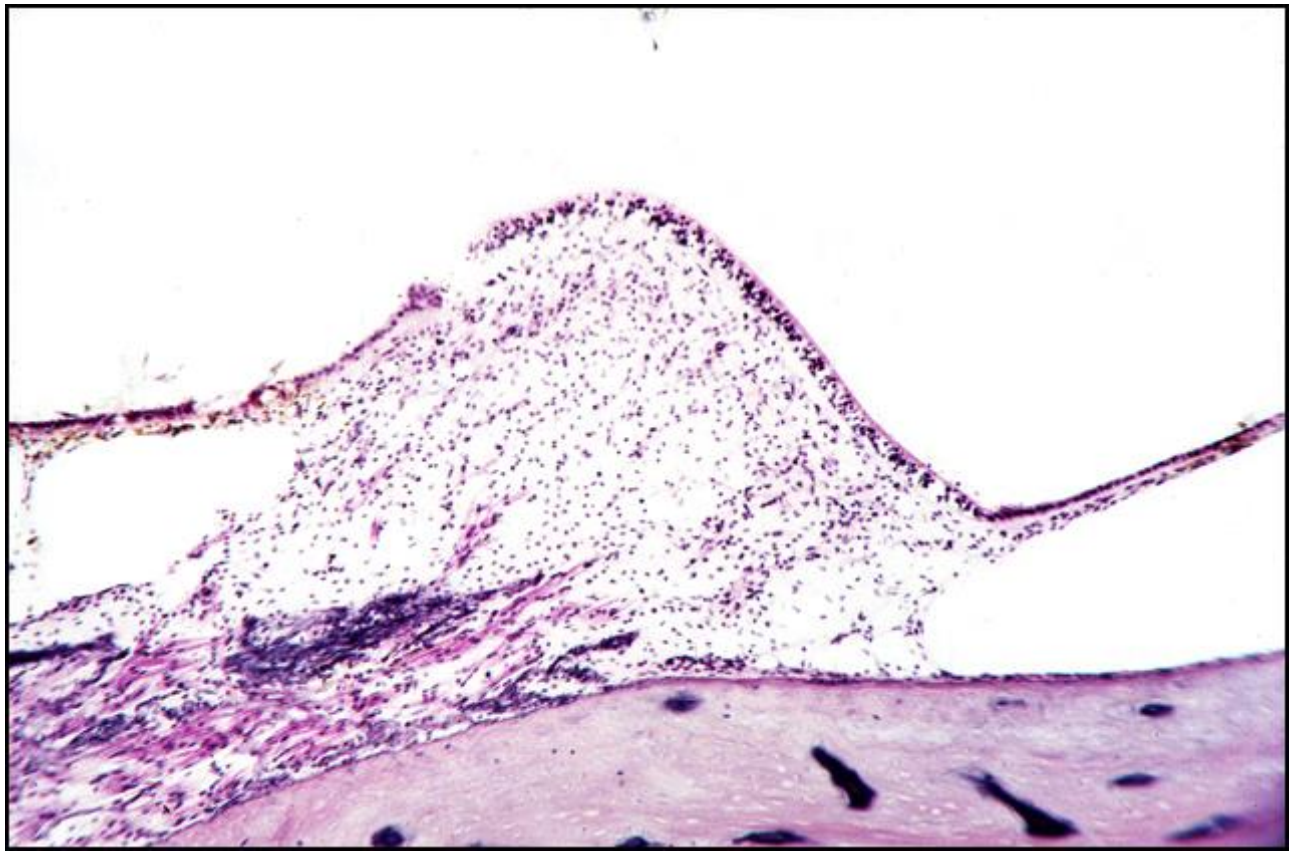
Comments: Lateral ampulla

Gender: Male

Age (yrs.): 56

Otologic Diagnosis:

1. Temporal bone, normal, left
2. Membranous labyrinth, normal, left
3. Bony labyrinth, normal, left
4. Artifact, removal, amputation, cochlea, partial, left
5. Artifact, manipulative intentional infracture of footplate, left
6. Artifact, bone dust, saw-g



DFNA9, L-61

Title: DFNA9, L-61

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 399

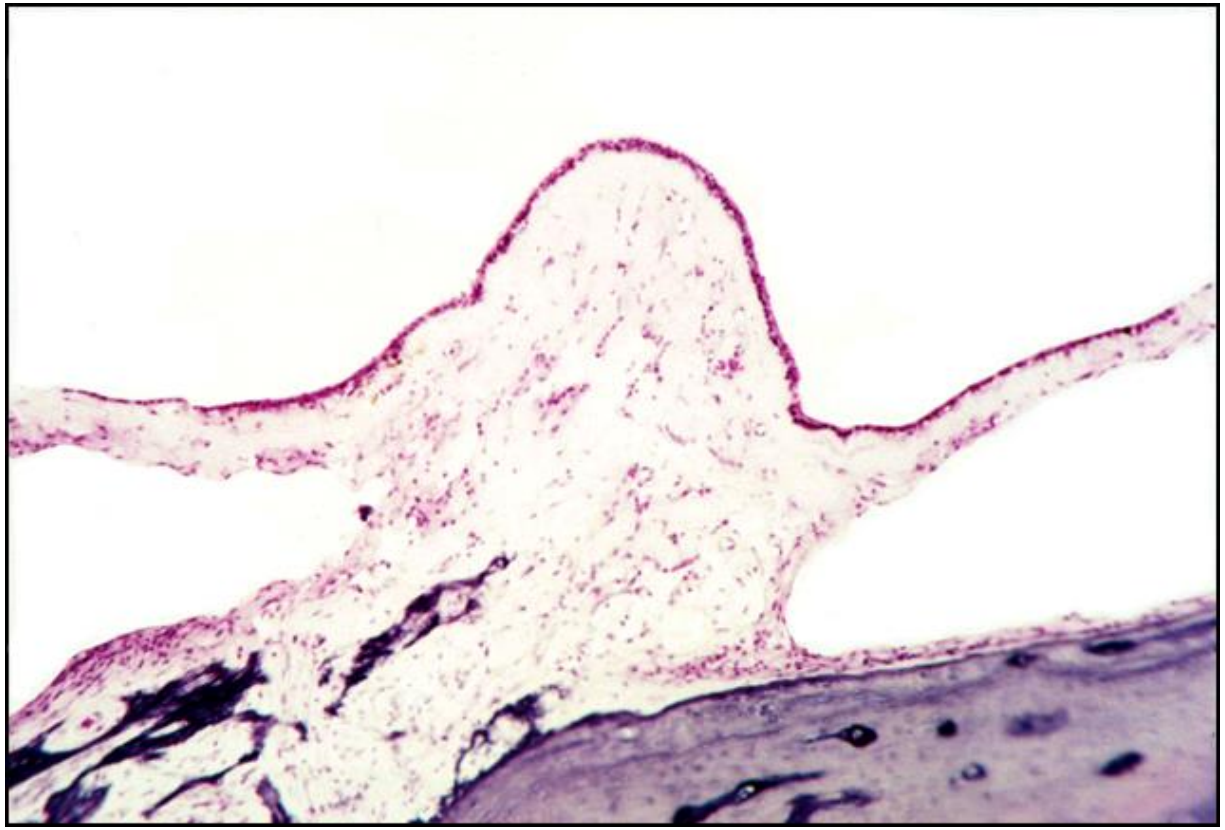
Comments: DFNA9 with COCH mutation, Lateral ampulla

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



DFNA9, R-201

Title: DFNA9, R-201

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 736

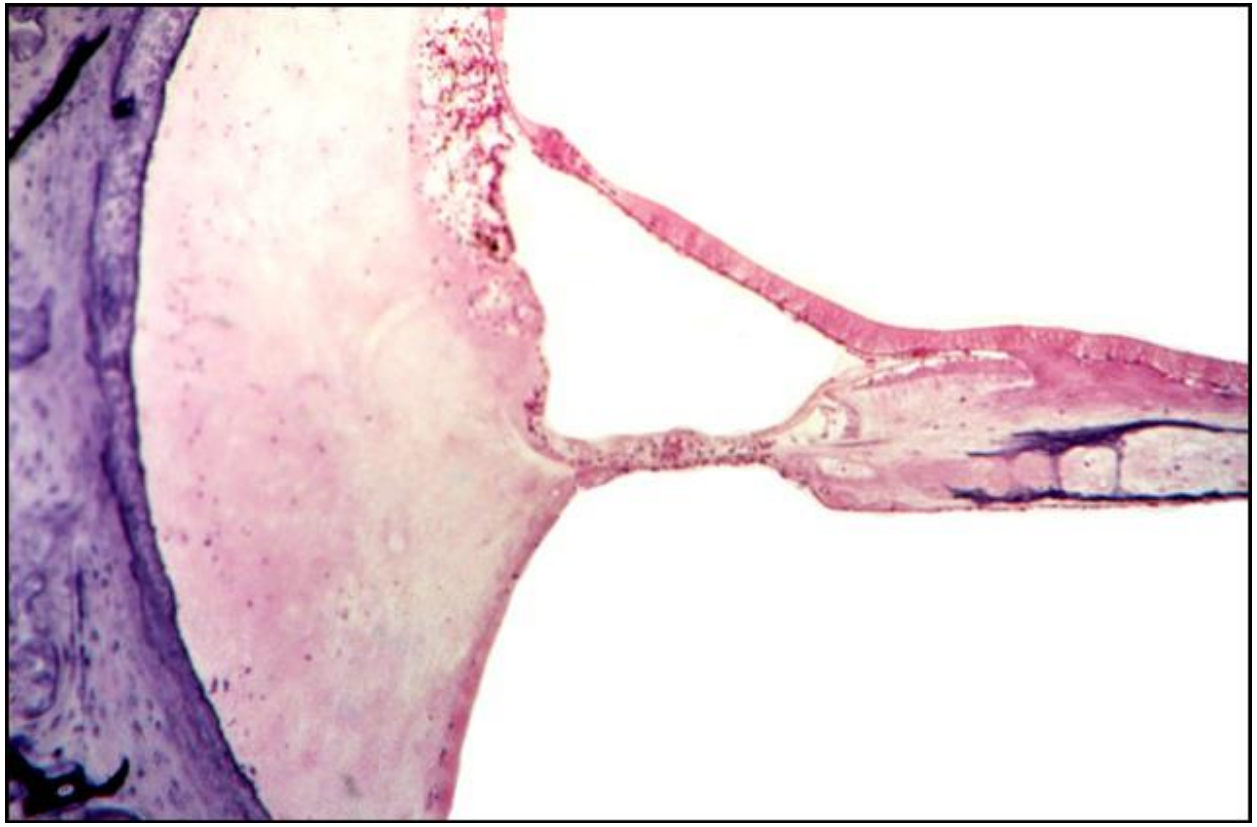
Comments: DFNA9 with COCH mutation, basal turn of cochlea

Gender: Female

Age (yrs.): 59

Otologic Diagnosis:

1. Hereditary sensorineural deafness, severe, bilateral
2. Proliferation of ground substance in supporting tissues of the auditory and vestibular sense organs with loss of fibrous tissue elements, bilateral
3. Loss of dendritic nerve fibers



DFNA9

Title: DFNA9

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 736

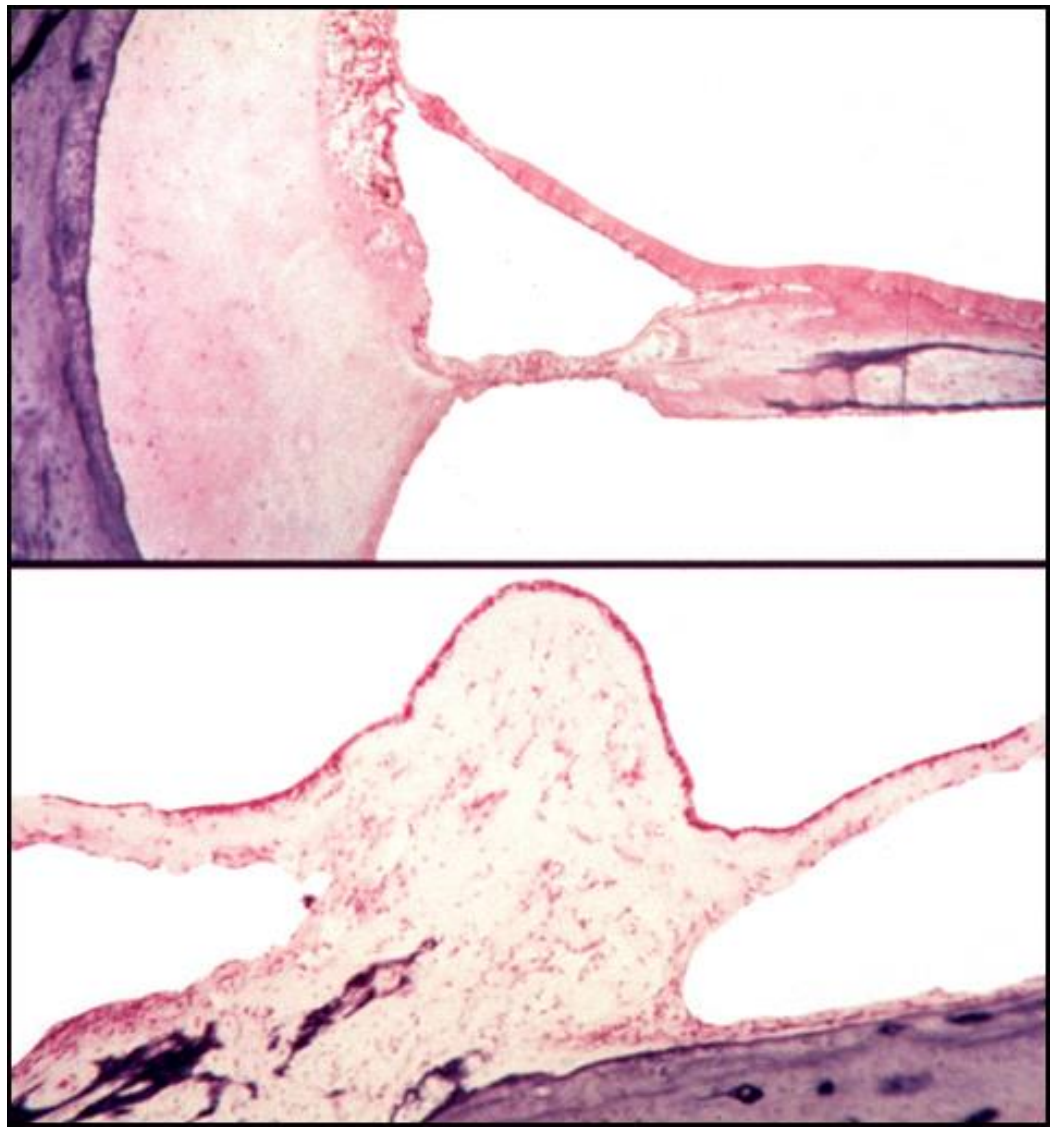
Comments: DFNA9 with COCH mutation

Gender: Female

Age (yrs.): 59

Otologic Diagnosis:

1. Hereditary sensorineural deafness, severe, bilateral
2. Proliferation of ground substance in supporting tissues of the auditory and vestibular sense organs with loss of fibrous tissue elements, bilateral
3. Loss of dendritic nerve fibers



DFNA9, L-209

Title: DFNA9, L-209

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

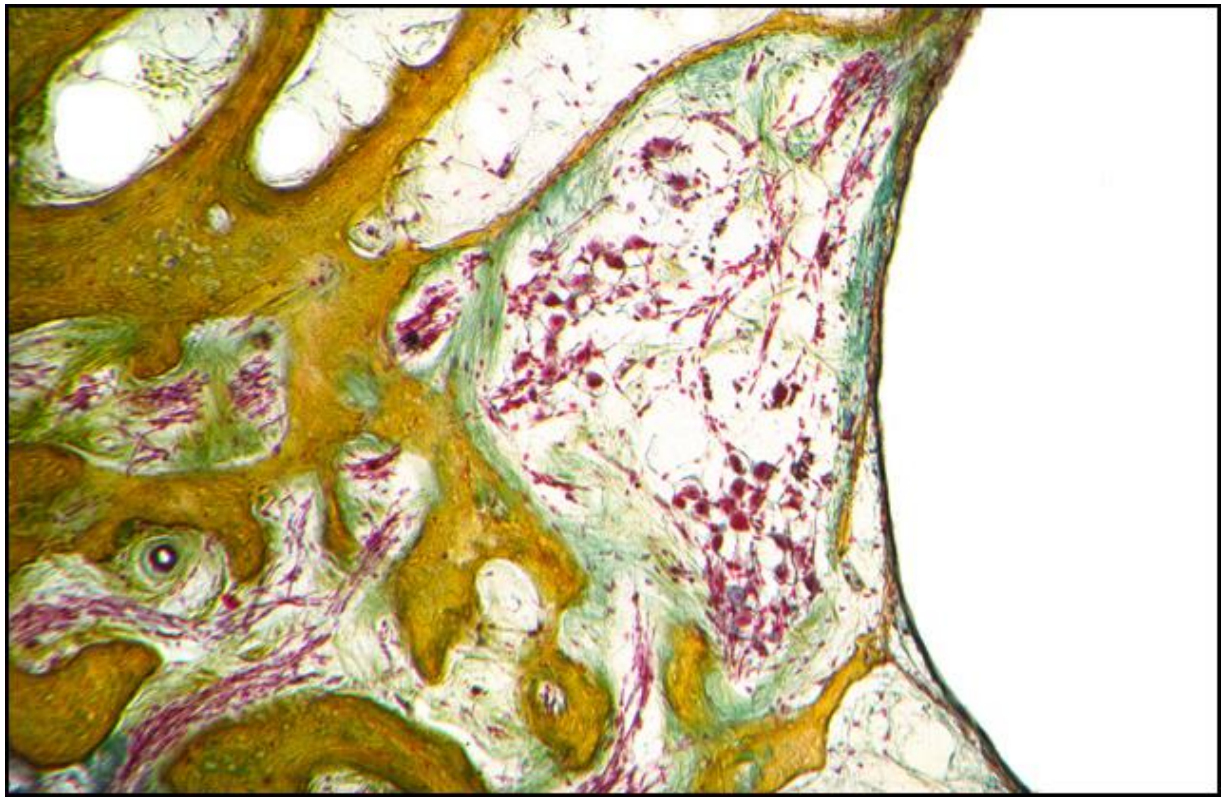
TB Case Number: 399

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



DFNA9, L-181

Title: DFNA9, L-181

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

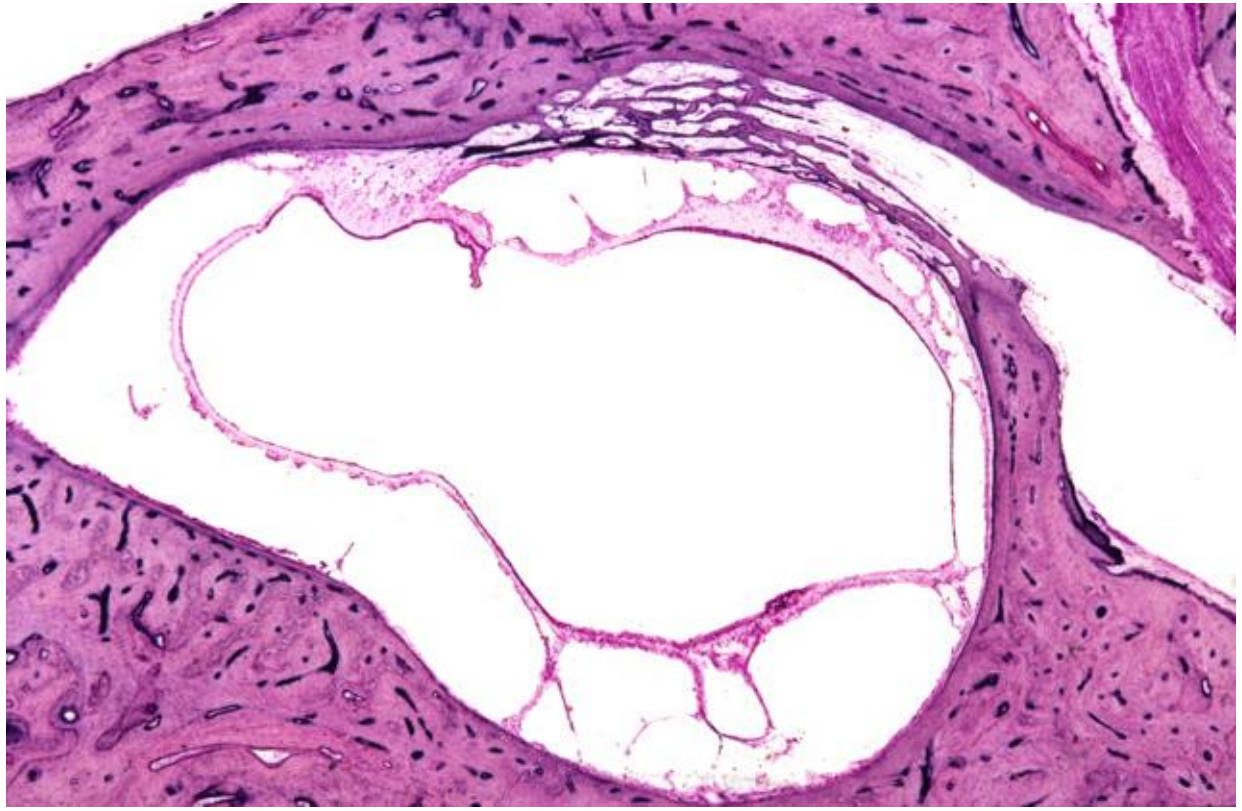
TB Case Number: 399

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



DFNA9, L-301

Title: DFNA9, L-301

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

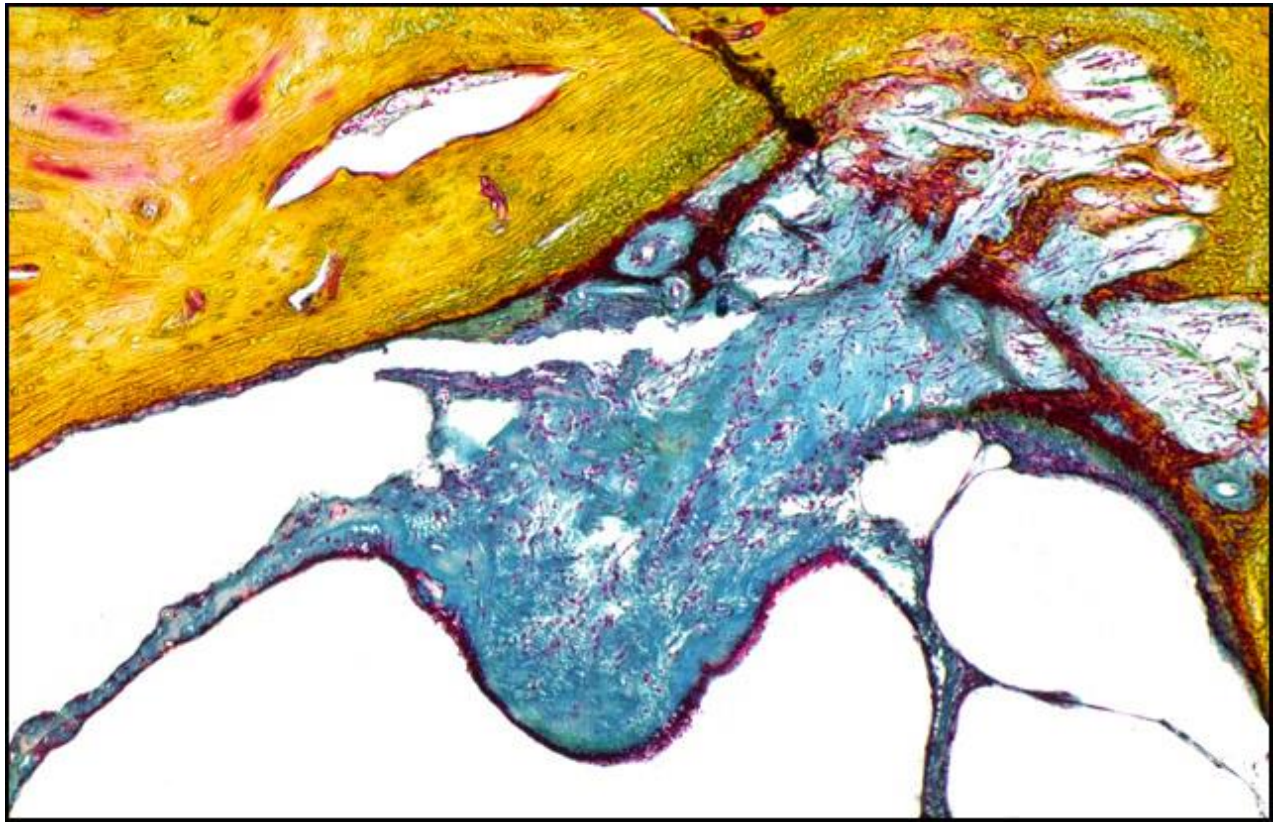
TB Case Number: 399

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



DFNA9, R-81

Title: DFNA9, R-81

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

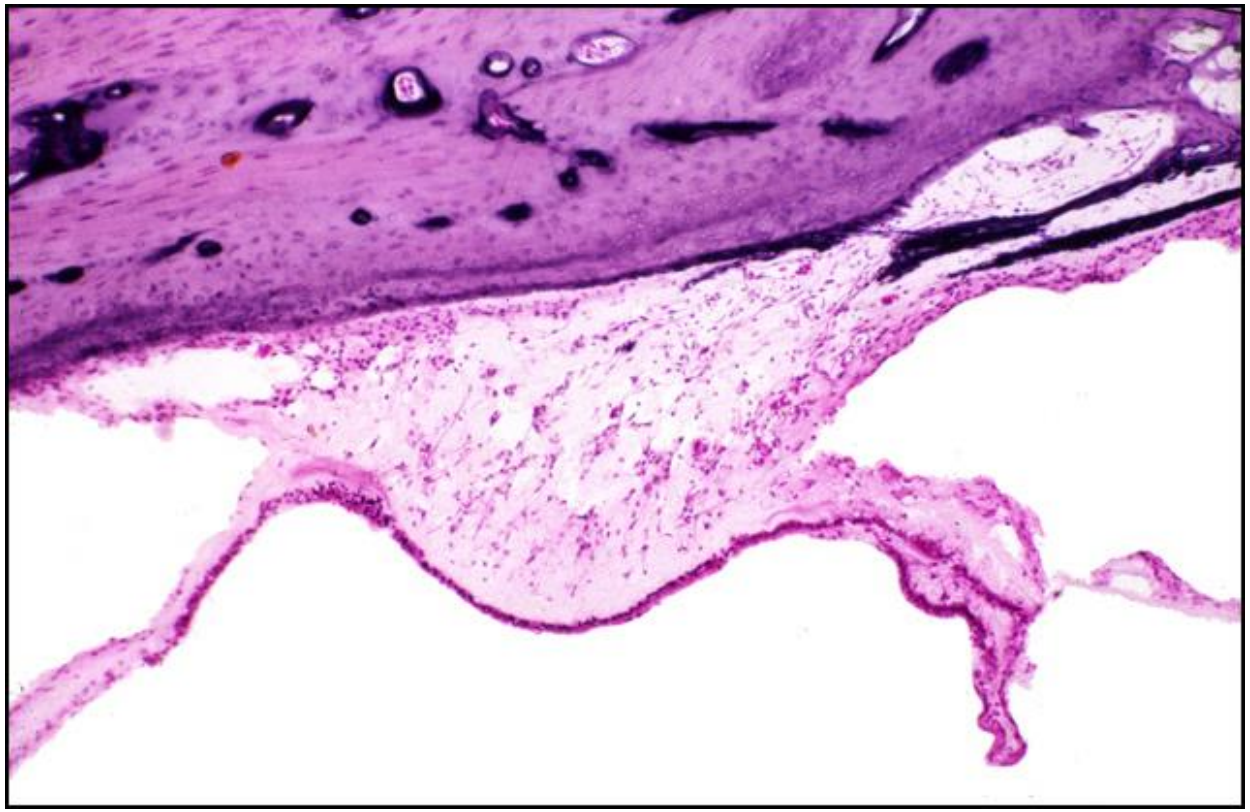
TB Case Number: 399

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



DFNA9, L-209

Title: DFNA9, L-209

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA 9

TB Case Number: 399

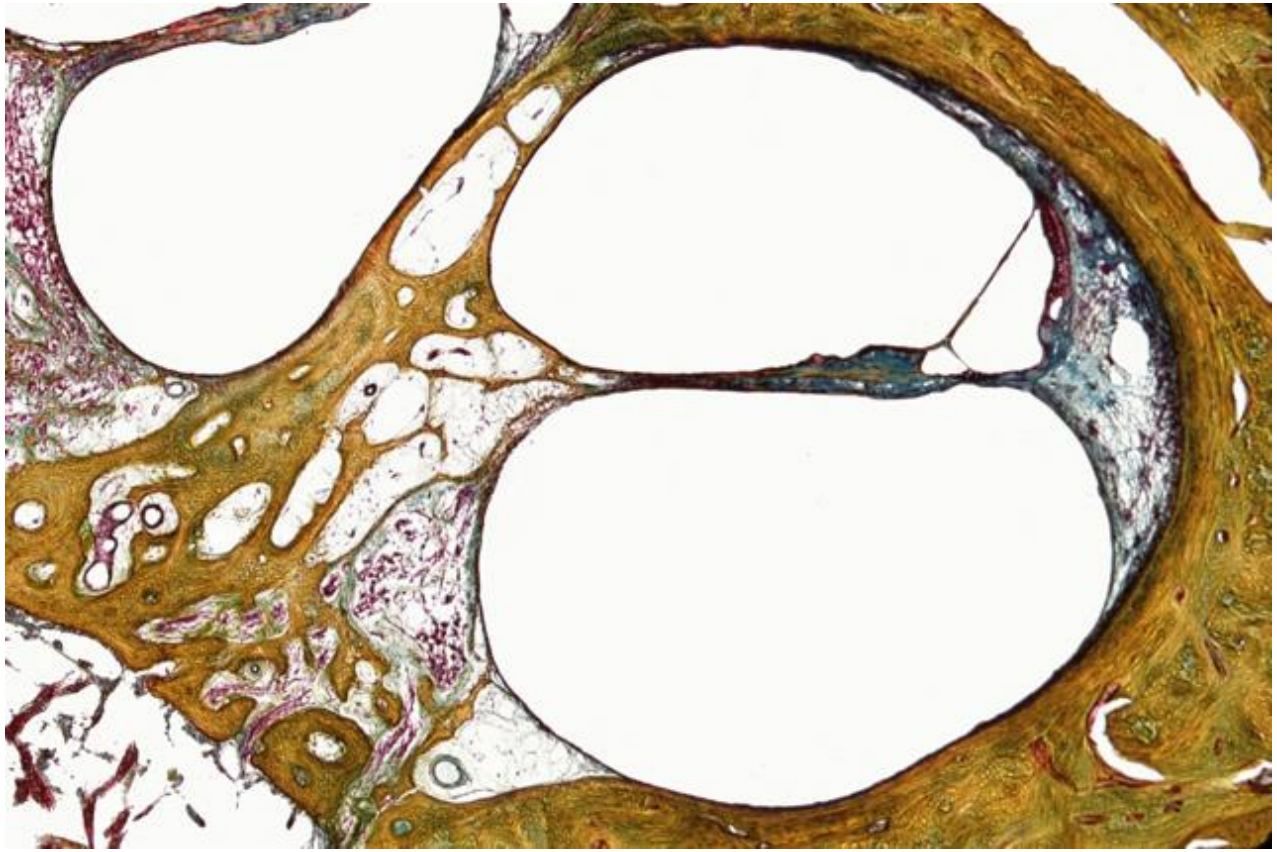
Comments: DFNA9 with COCH mutation, Movat's pentachrome stain

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



DFNA9, L-209

Title: DFNA9, L-209

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 399

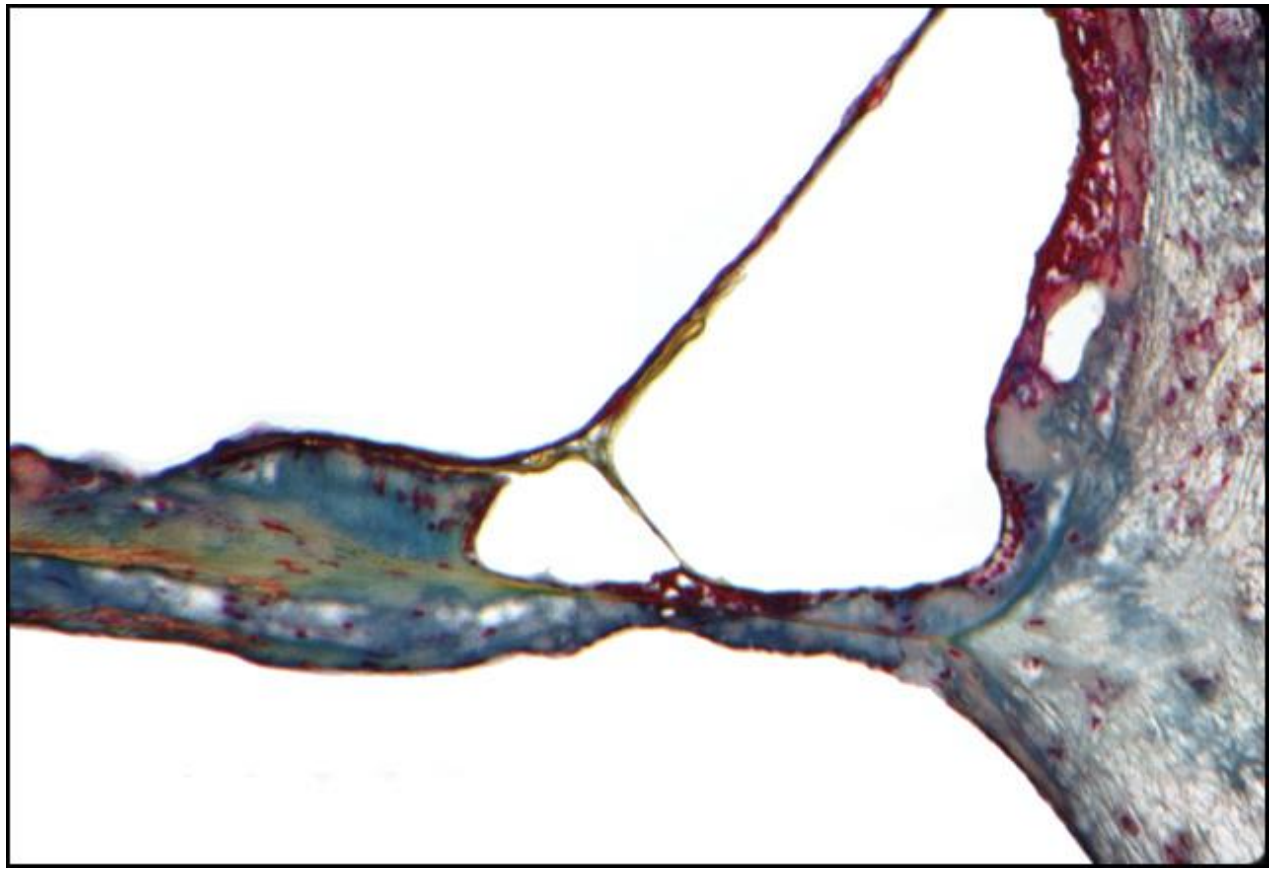
Comments: DFNA9 with COCH mutation, Movat's pentachrome stain

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



DFNA9, L-209

Title: DFNA9, L-209

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 399

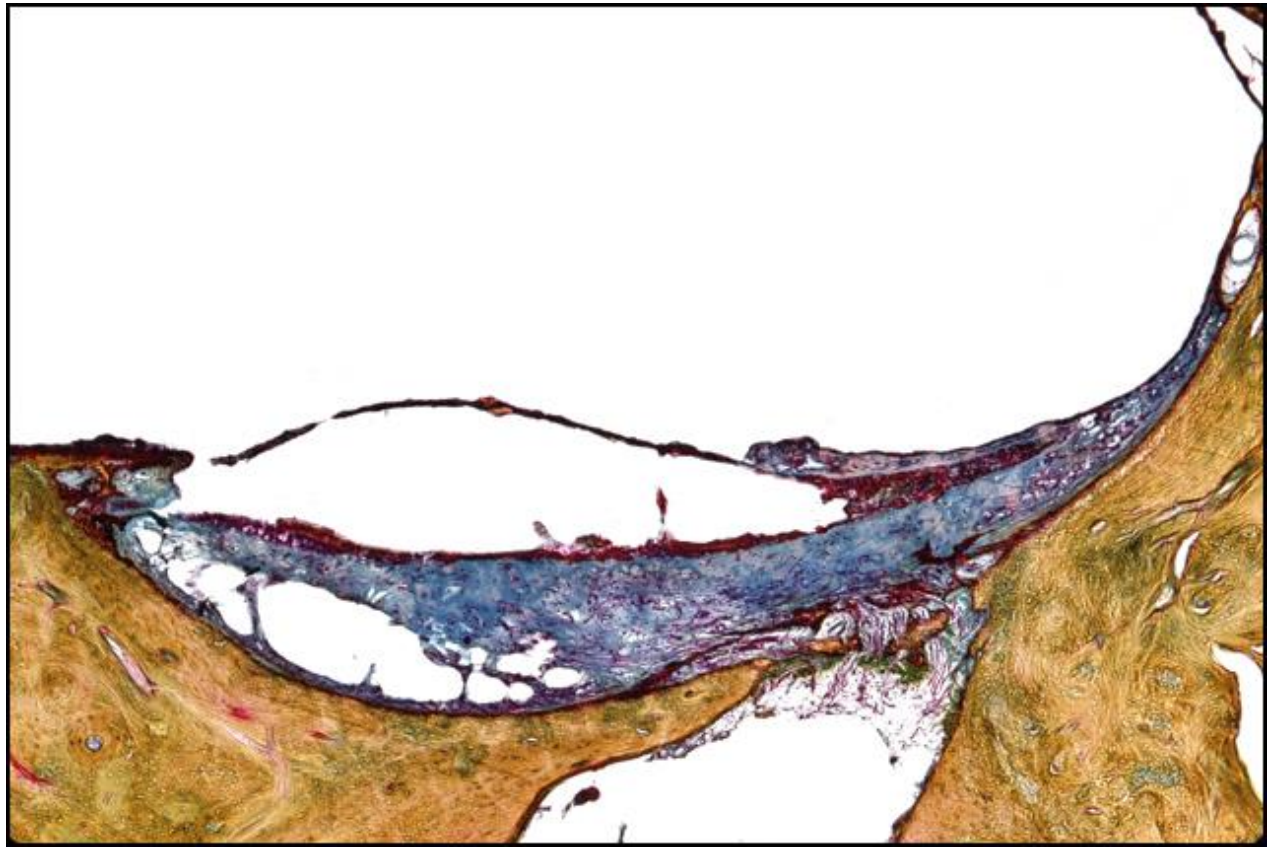
Comments: DFNA9 with COCH mutation, Movat's pentachrome stain, saccule

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



DFNA9, L-209

Title: DFNA9, L-209

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 399

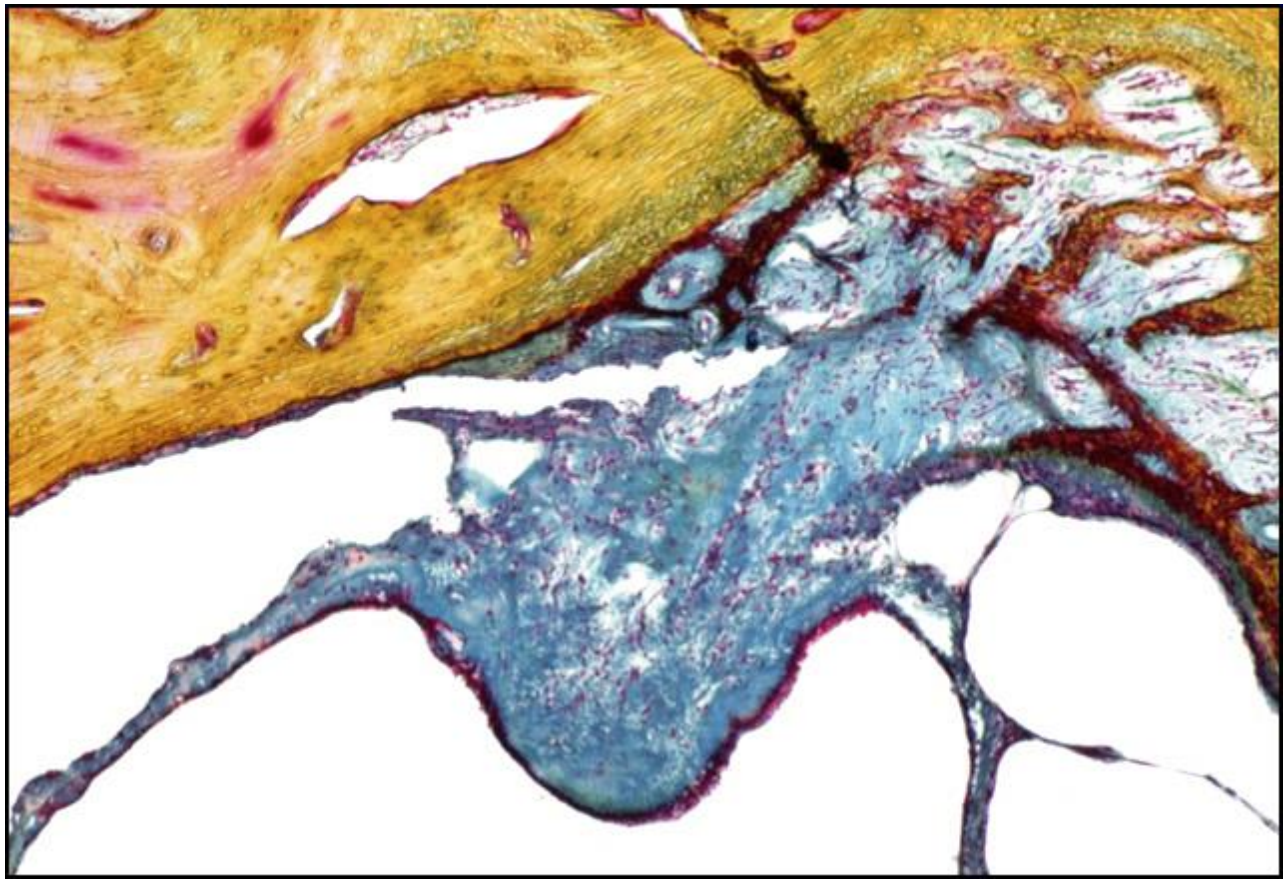
Comments: DFNA9 with COCH mutation, Movat's pentachrome stain, Posterior ampulla

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Arachnoid cyst, internal auditory canal
2. Degeneration, vestibular sense organs, severe
3. Atrophic degenerative changes in structures of the cochlear duct
4. Severe degeneration, vestibular nerves
5. Degeneration of the cochlear nerve
6. Otosclerosis



Unaffected, L-247

Title: Unaffected, L-247

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 733

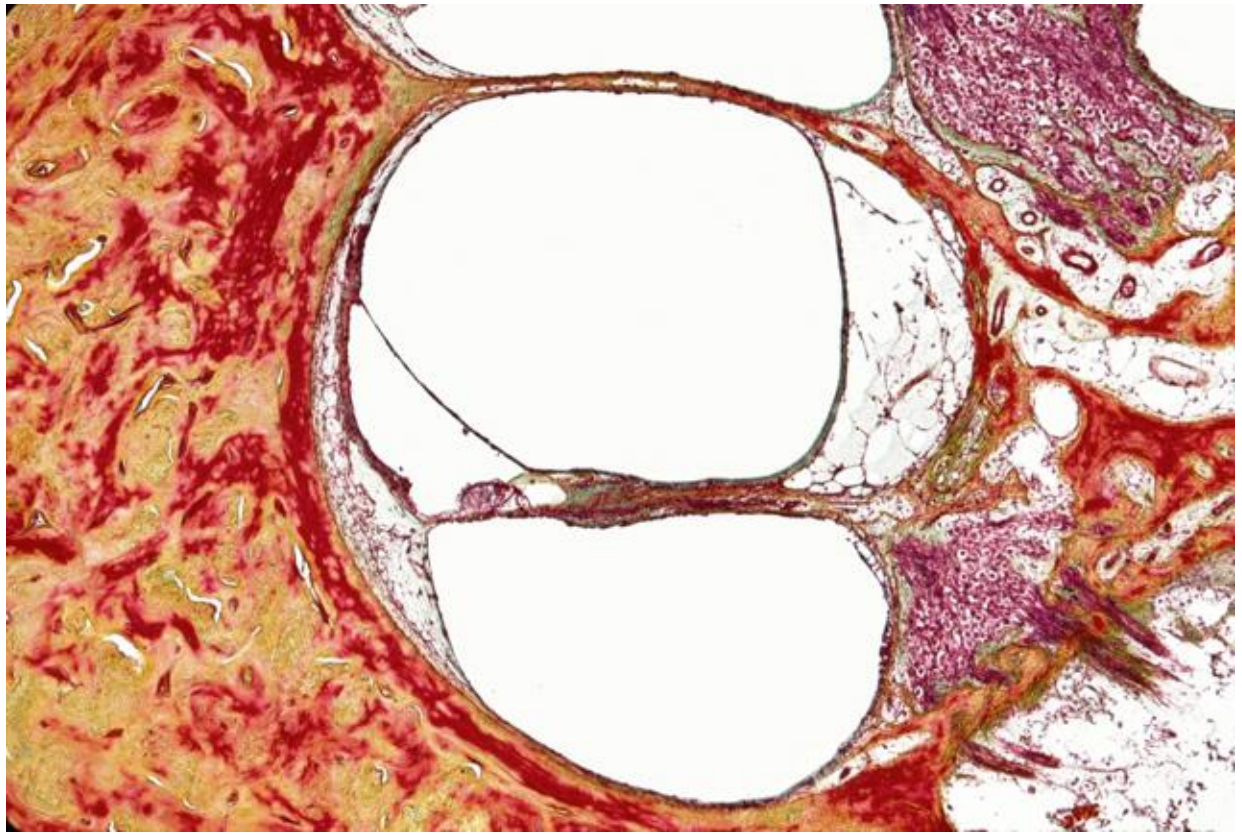
Comments: Movat's pentachrome stain

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Sensorineural hearing loss due to aging, combination strial atrophy and inner ear conductive lesion
2. Acoustic trauma, bilateral, mild
3. Excellent anatomical preparation



Unaffected, L-247

Title: Unaffected, L-247

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 733

Comments: Movat's pentachrome stain

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Sensorineural hearing loss due to aging, combination strial atrophy and inner ear conductive lesion
2. Acoustic trauma, bilateral, mild
3. Excellent anatomical preparation

Unaffected, L-247

Title: Unaffected, L-247

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 733

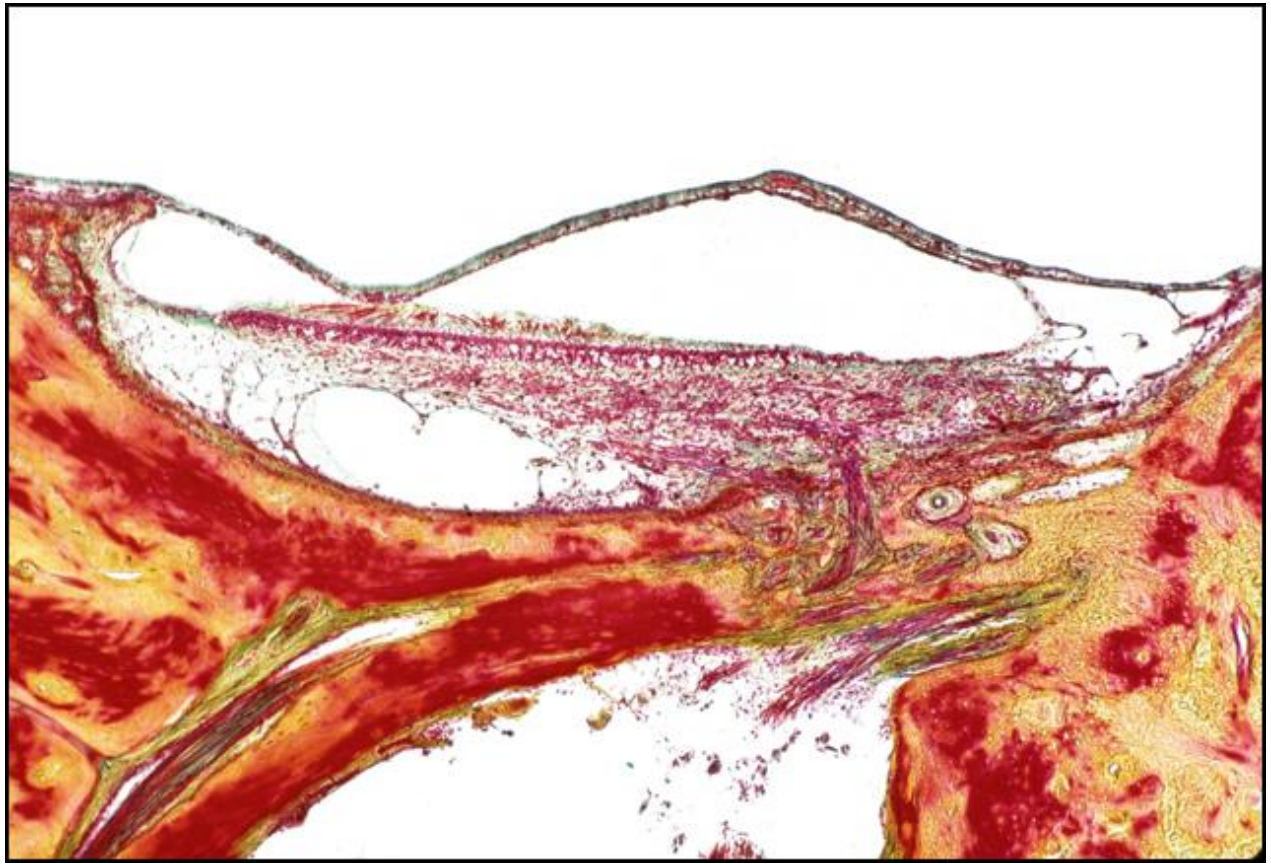
Comments: Movat's pentachrome stain, saccule

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Sensorineural hearing loss due to aging, combination strial atrophy and inner ear conductive lesion
2. Acoustic trauma, bilateral, mild
3. Excellent anatomical preparation



Unaffected, R-303

Title: Unaffected, R-303

Chapter: Developmental Defects

Chapter Section: Non-Syndromic Hearing Loss / DFNA9

TB Case Number: 733

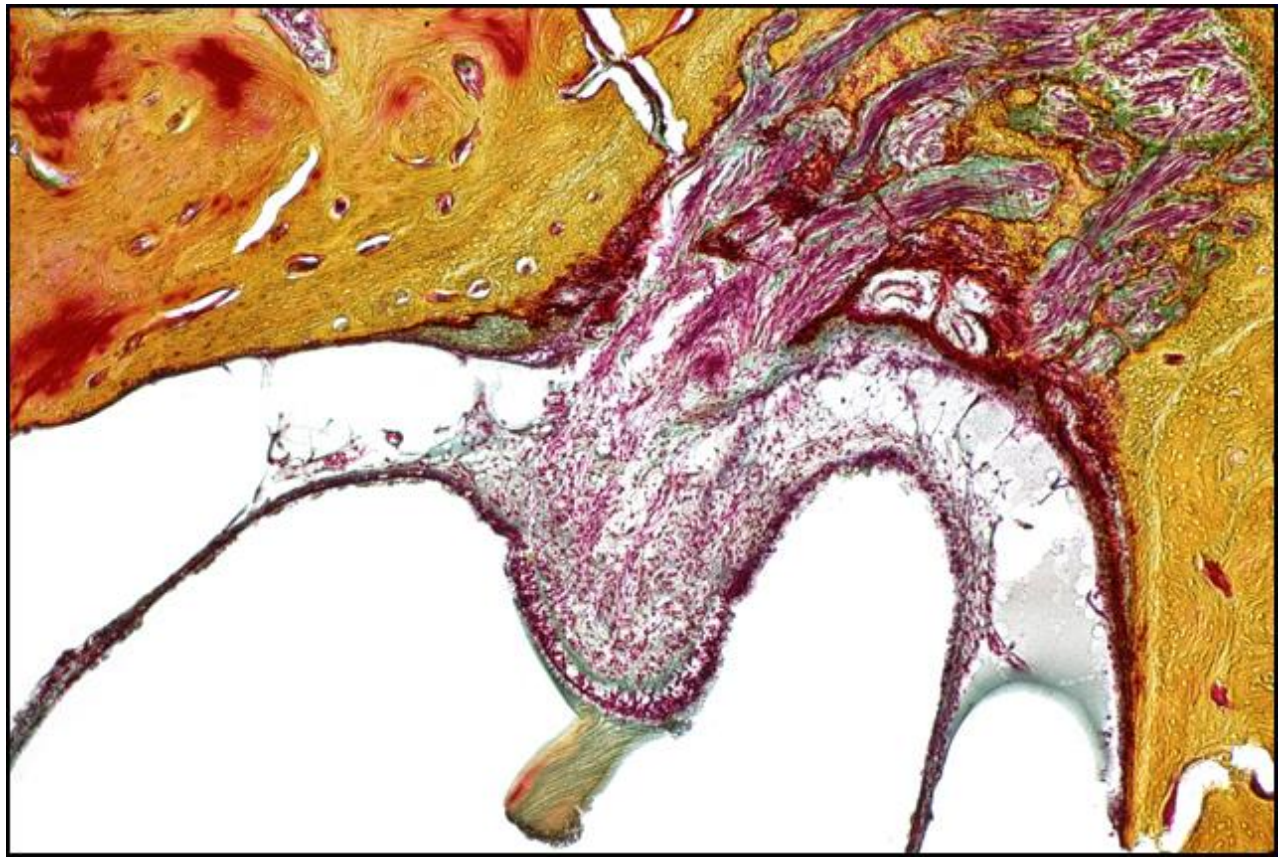
Comments: Movat's pentachrome stain, Posterior ampulla

Gender: Male

Age (yrs.): 81

Otologic Diagnosis:

1. Sensorineural hearing loss due to aging, combination strial atrophy and inner ear conductive lesion
2. Acoustic trauma, bilateral, mild
3. Excellent anatomical preparation



Waardenburg Type I, R-91

Title: Waardenburg Type I, R-91

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

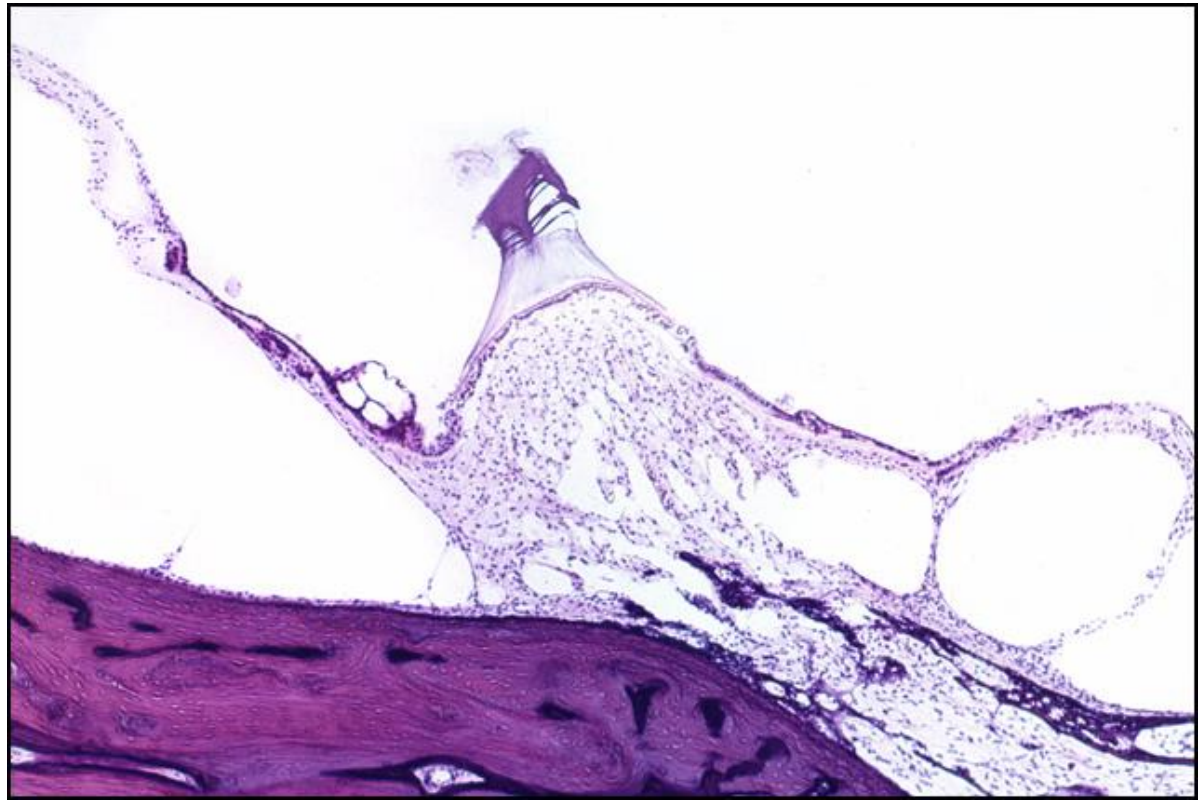
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, L-81

Title: Waardenburg Type I, L-81

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

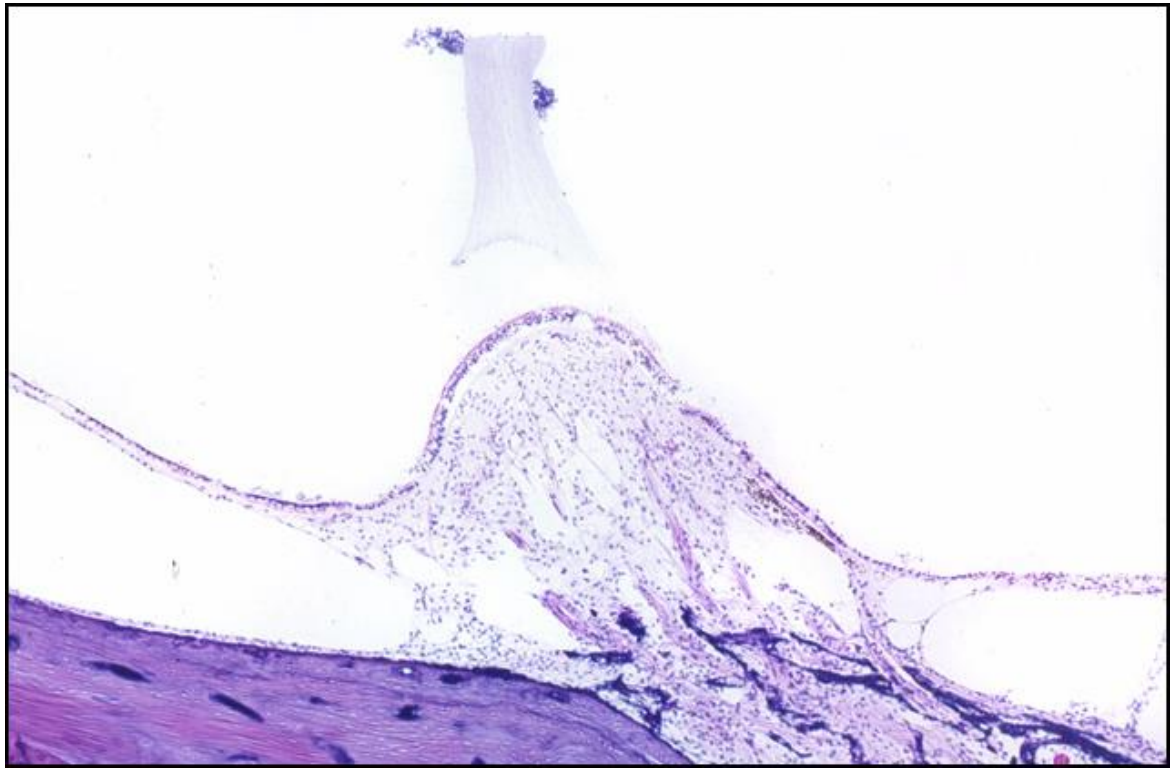
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, R-181

Title: Waardenburg Type I, R-181

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

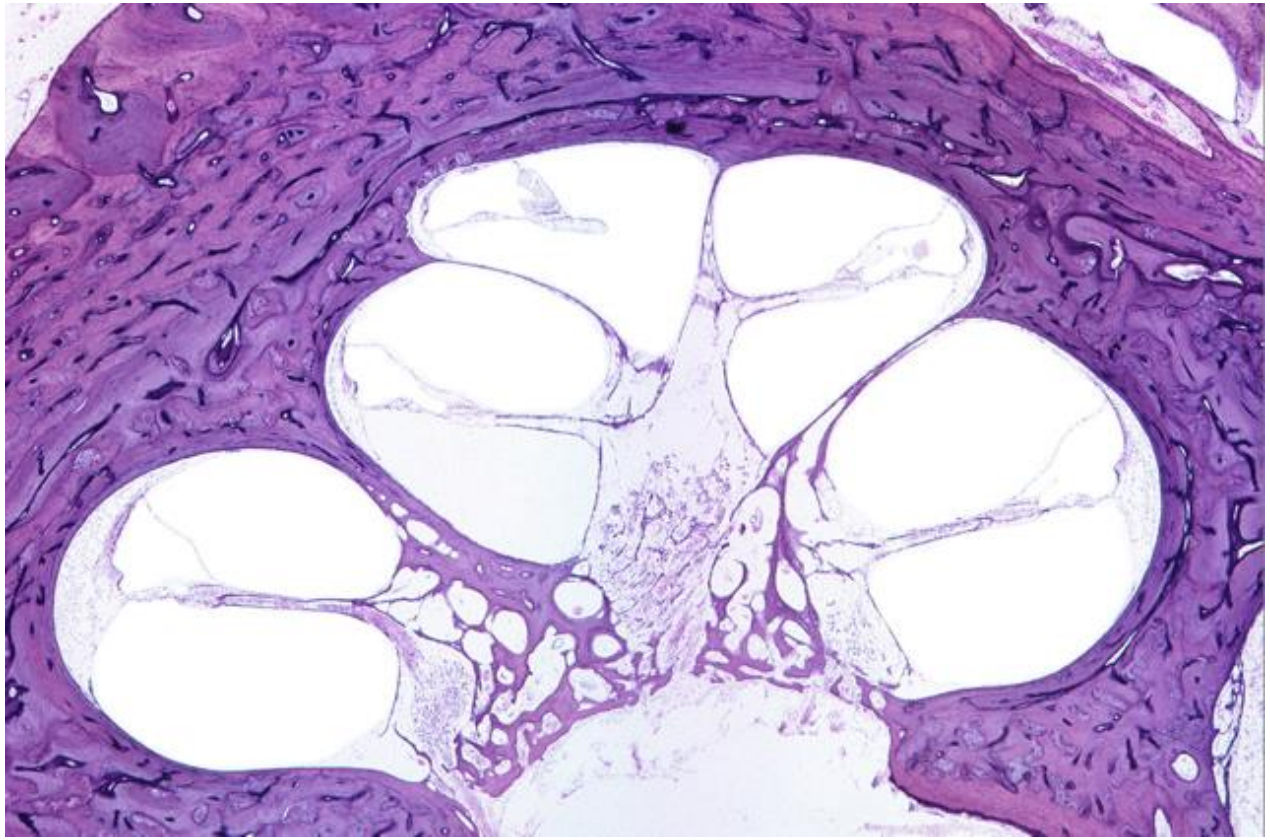
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, L-201

Title: Waardenburg Type I, L-201

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

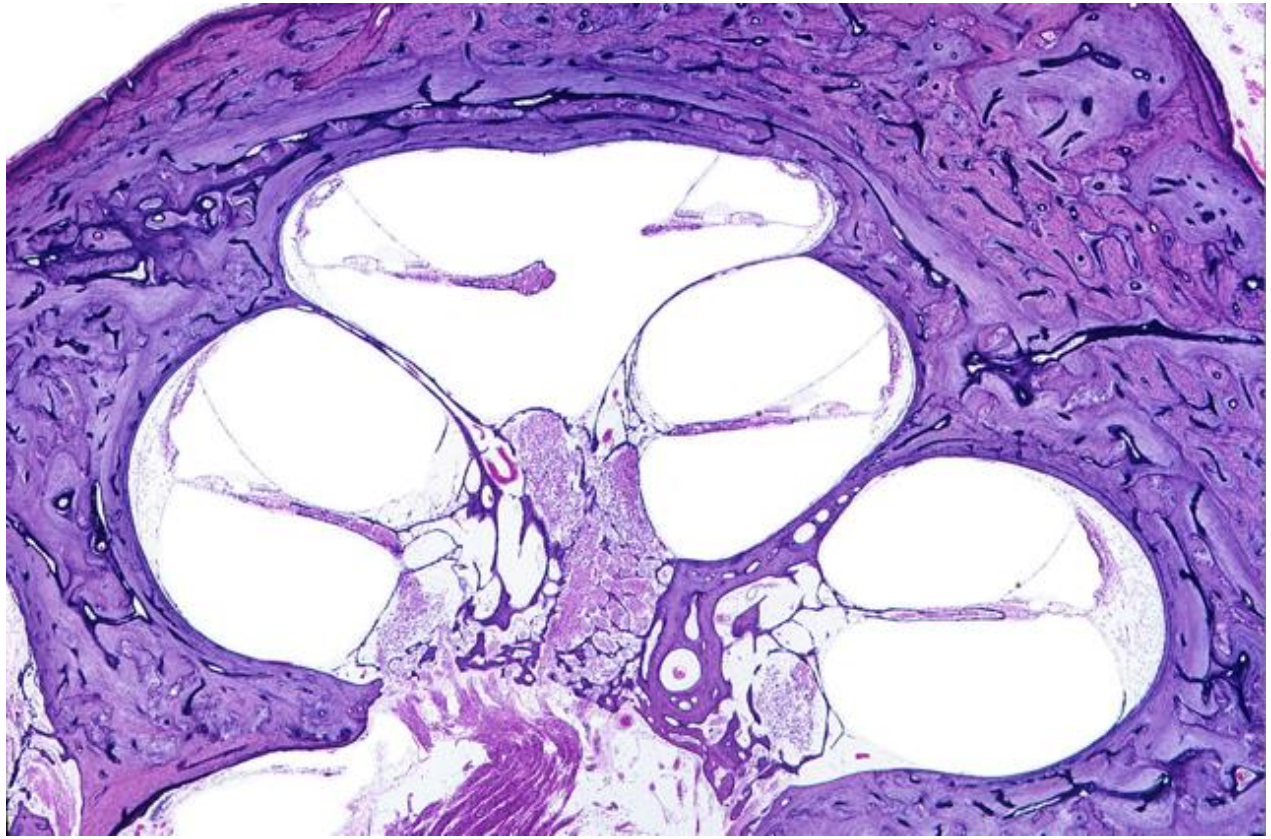
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, R-91

Title: Waardenburg Type I, R-91

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

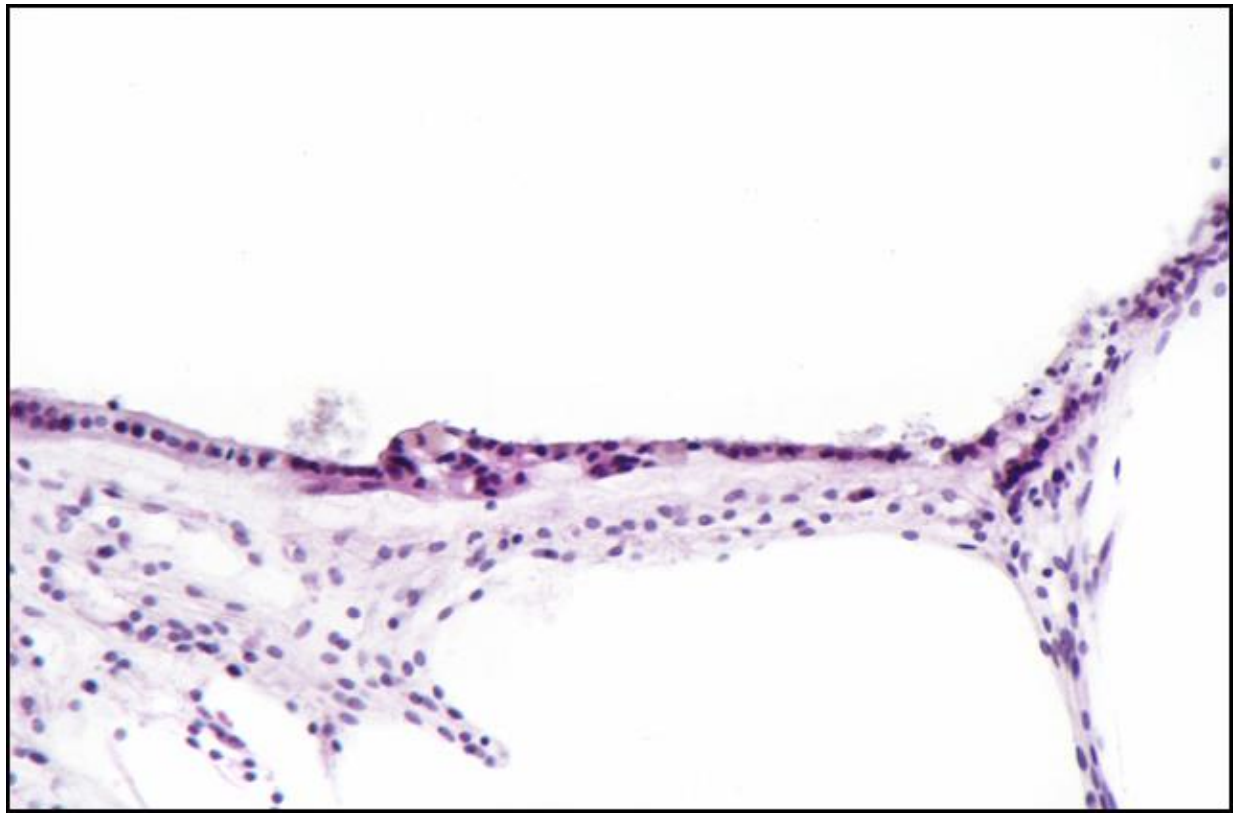
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, L-81

Title: Waardenburg Type I, L-81

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

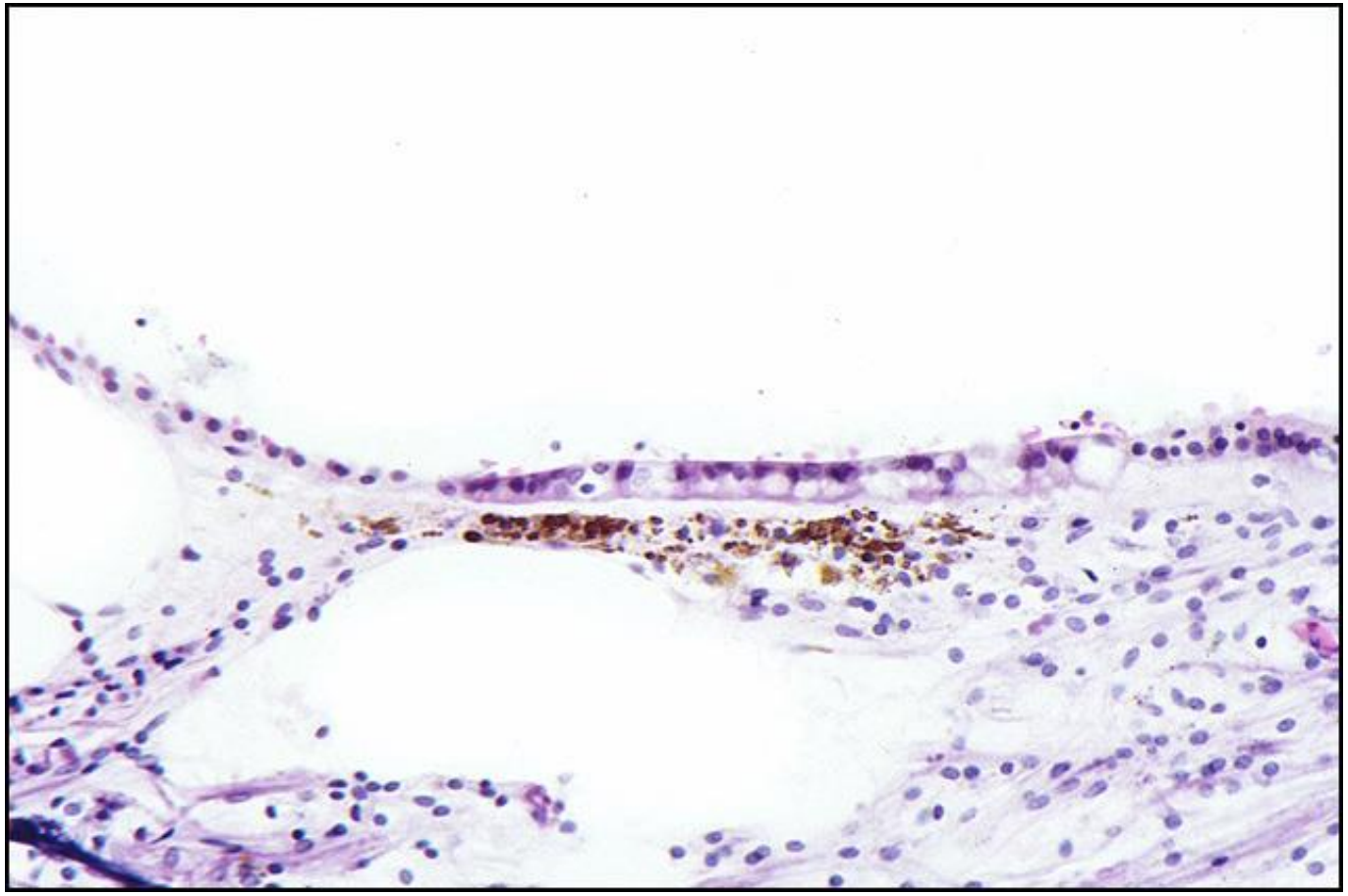
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, R-181

Title: Waardenburg Type I, R-181

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

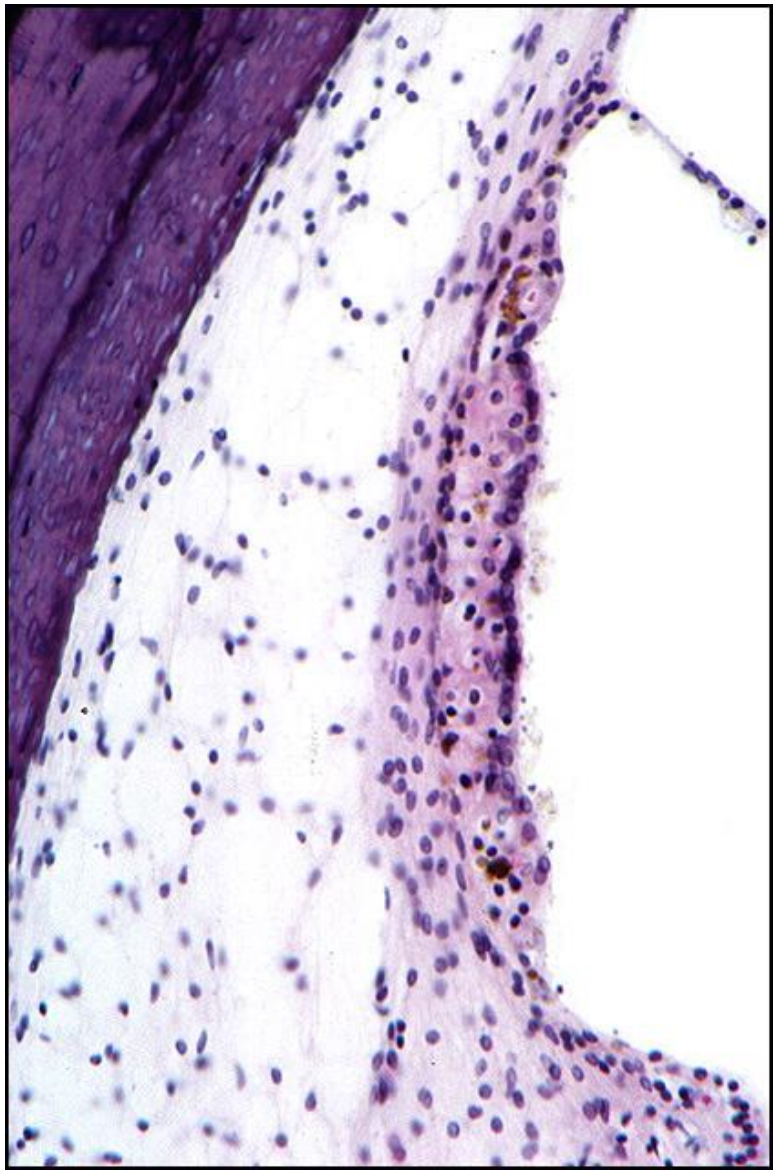
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, L-201

Title: Waardenburg Type I, L-201

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

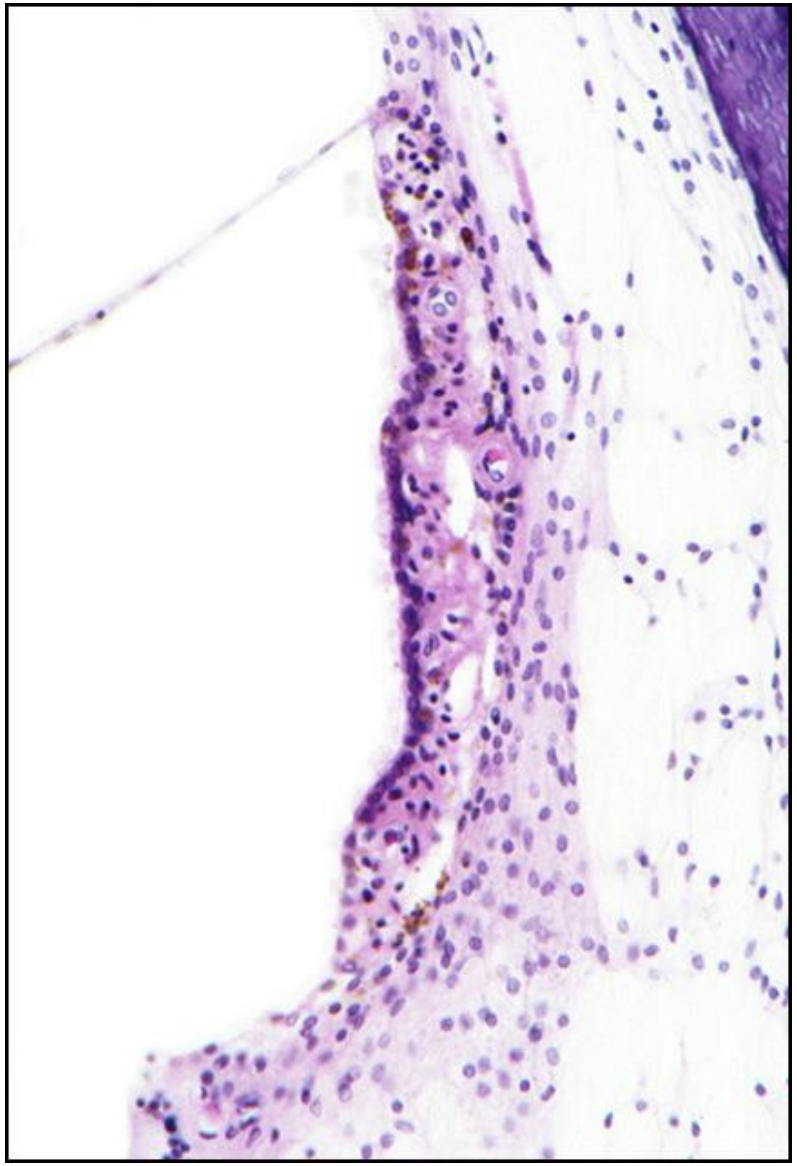
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I

Title: Waardenburg Type I

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

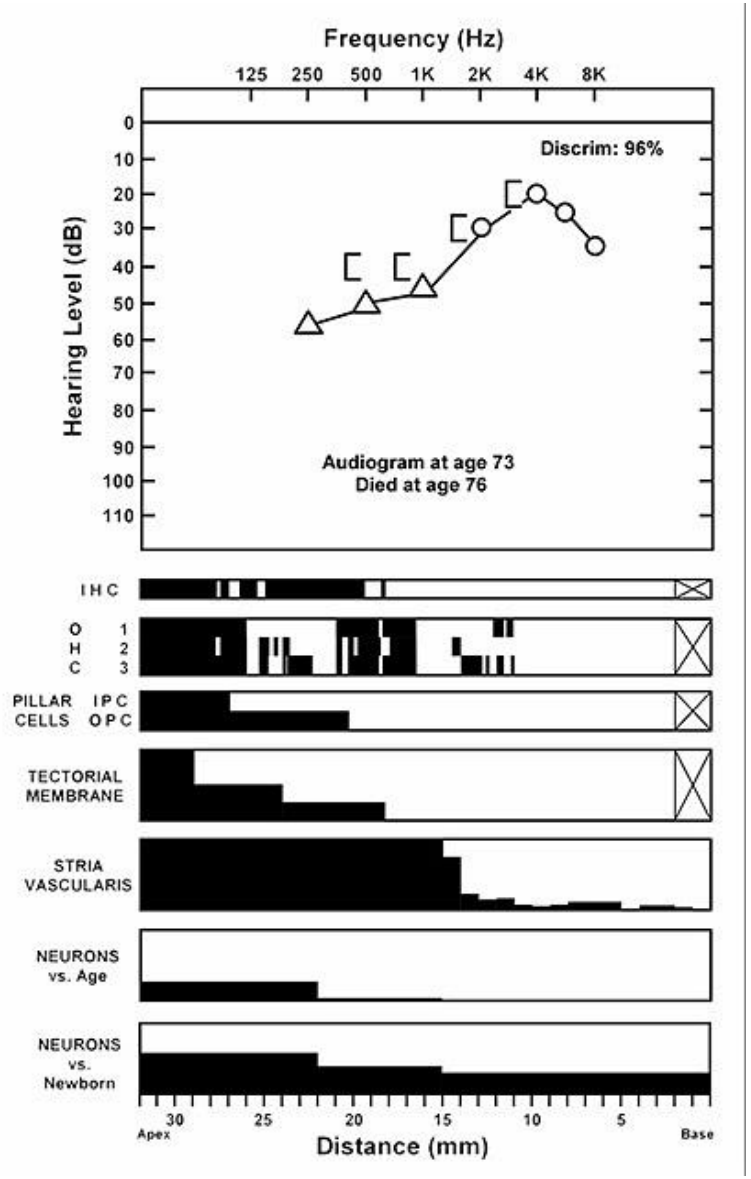
Comments: Waardenburg Type I, Audio-cytocochleogram

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I

Title: Waardenburg Type I

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

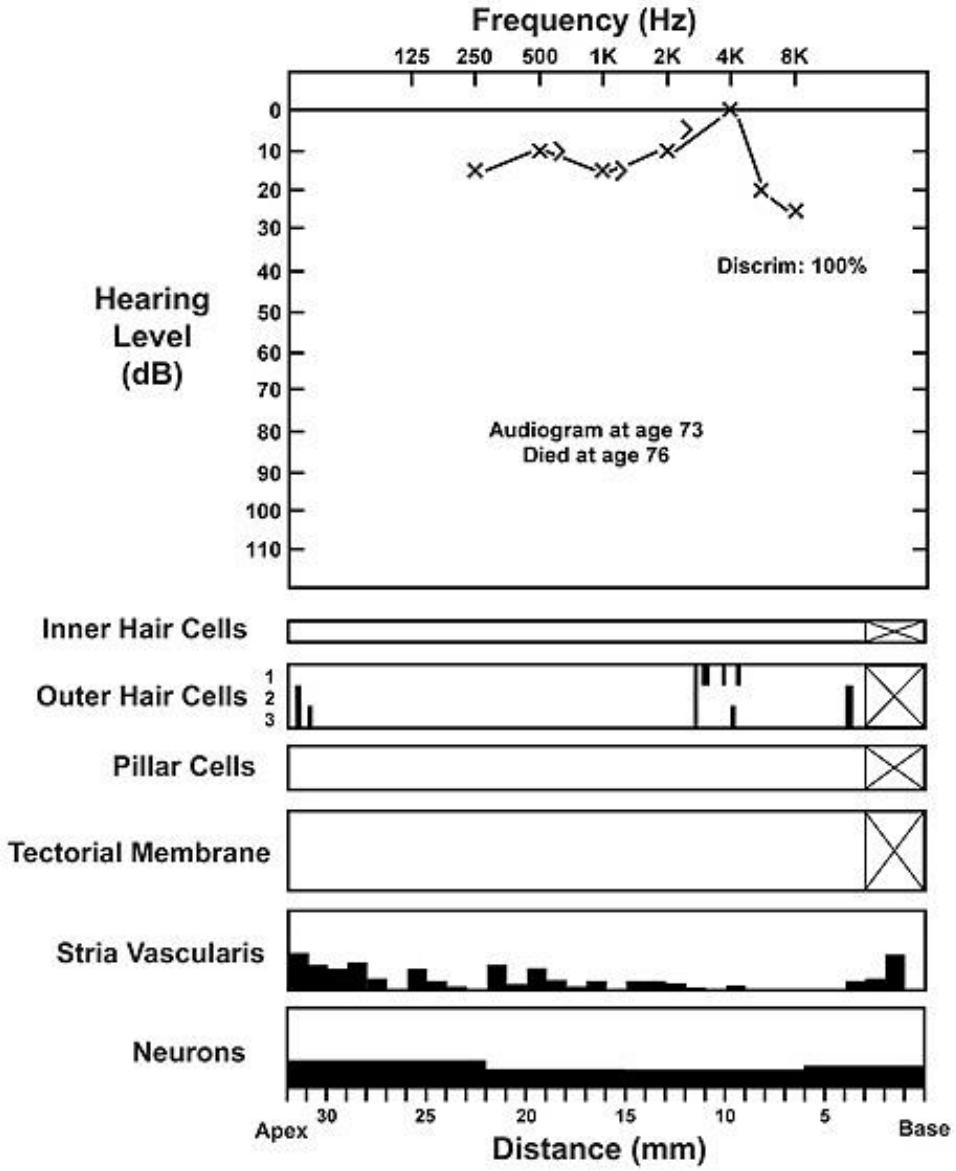
Comments: Waardenburg Type I, Audio-cytocochleogram

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, R-181

Title: Waardenburg Type I, R-181

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

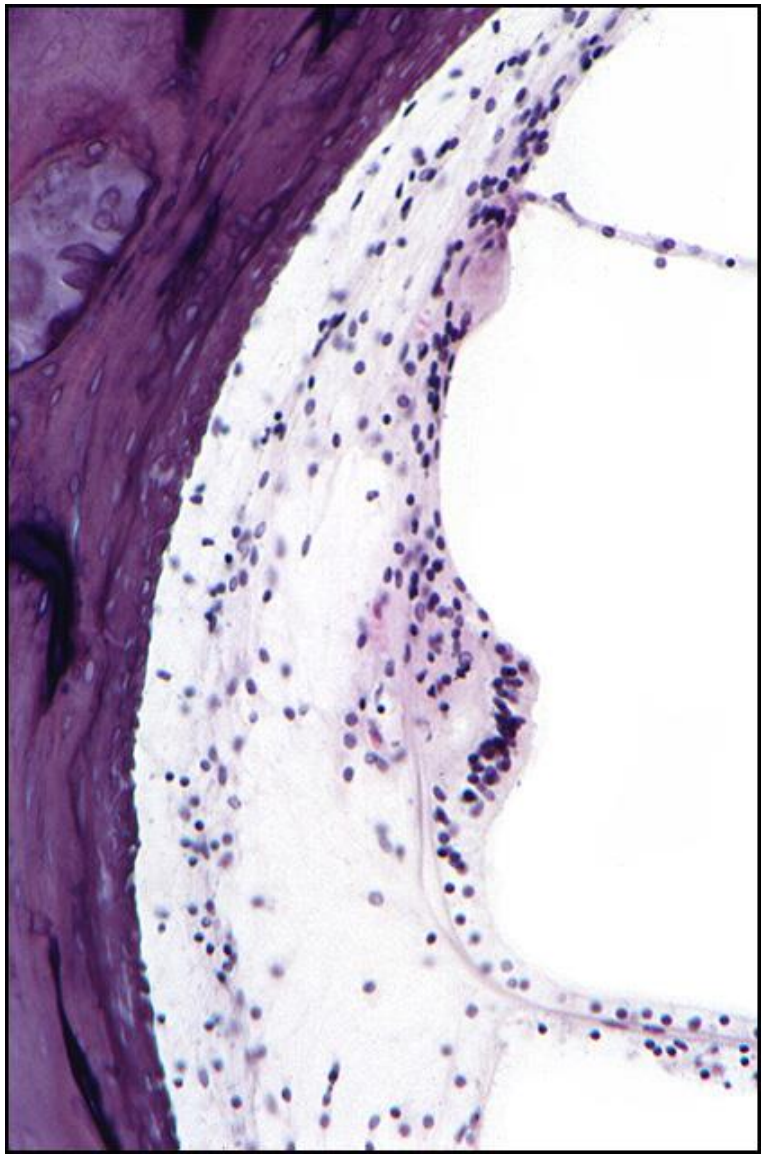
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, L-201

Title: Waardenburg Type I, L-201

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

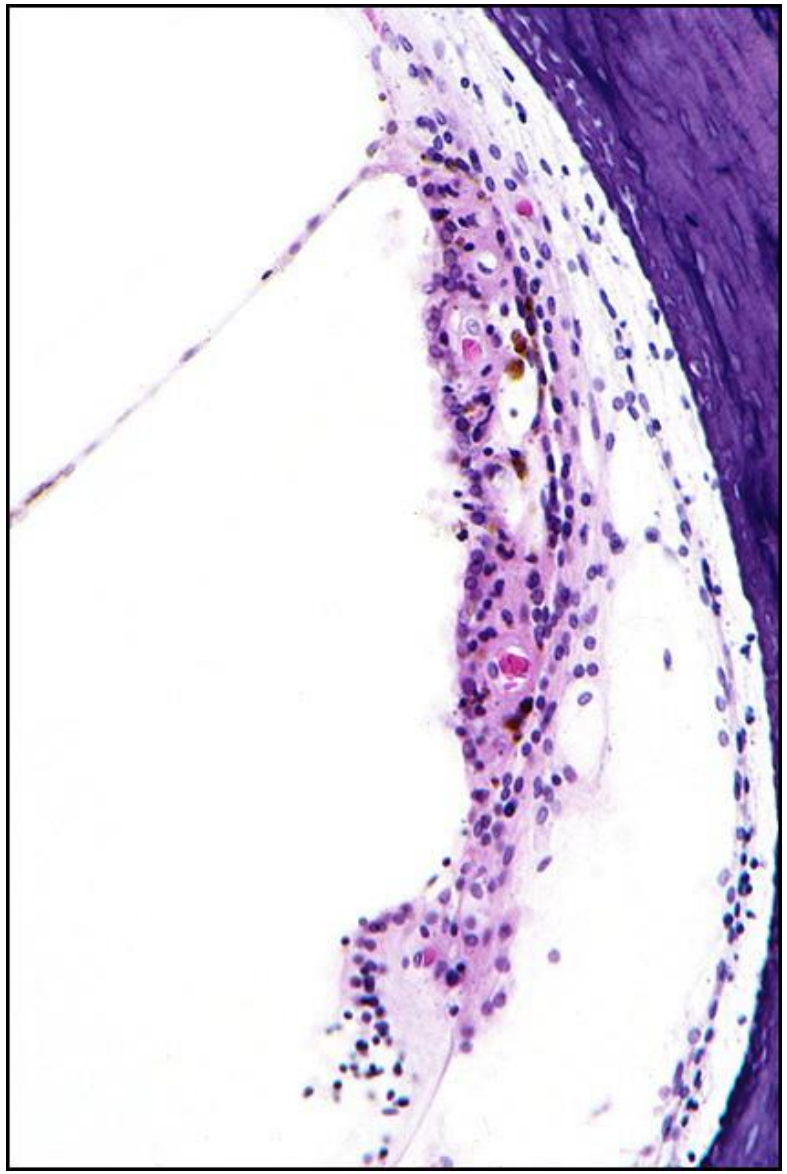
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, R-181

Title: Waardenburg Type I, R-181

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

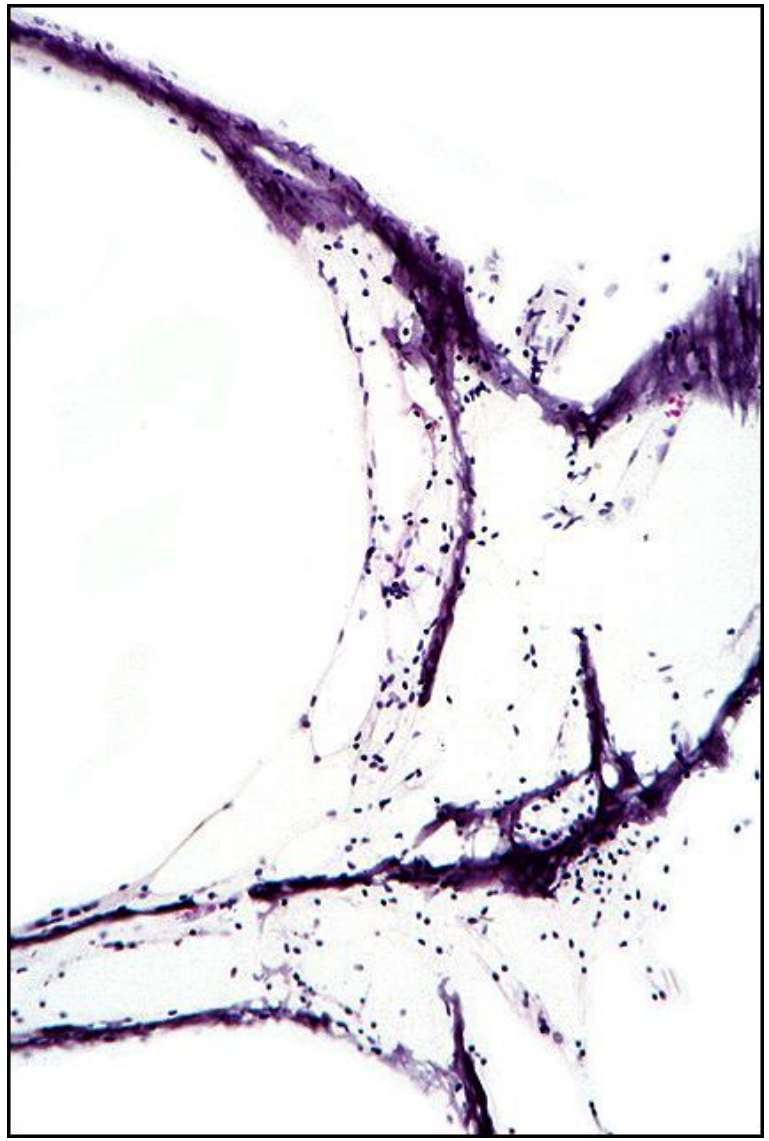
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Waardenburg Type I, L-201

Title: Waardenburg Type I, L-201

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Waardenburg Syndrome

TB Case Number: 907

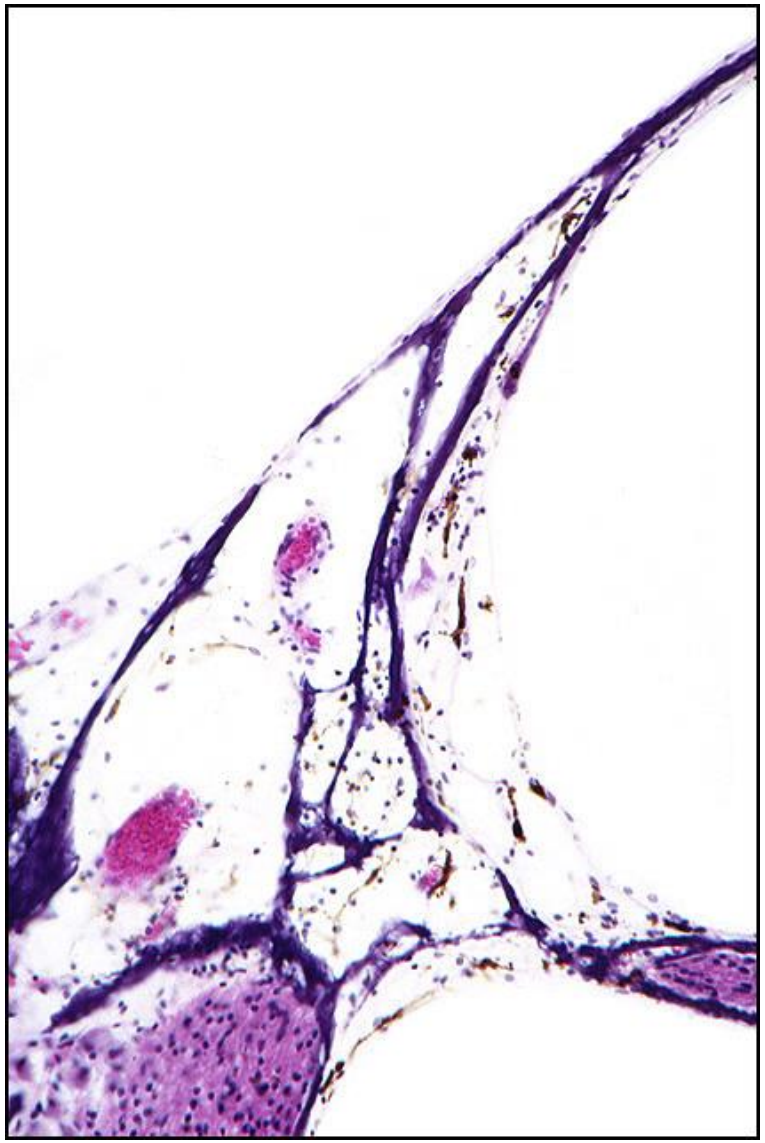
Comments: Waardenburg Type I

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Waardenburg's syndrome, type I, with sensorineural hearing loss, right
2. Dysmorphogenesis, cochlea and vestibular labyrinth, due to Waardenburg's syndrome, right
3. Jugular bulb, high, left



Mohr-Tranebjaerg Syndrome

Title: Mohr-Tranebjaerg Syndrome

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mohr-Tranebjaerg Syndrome

TB Case Number: 920

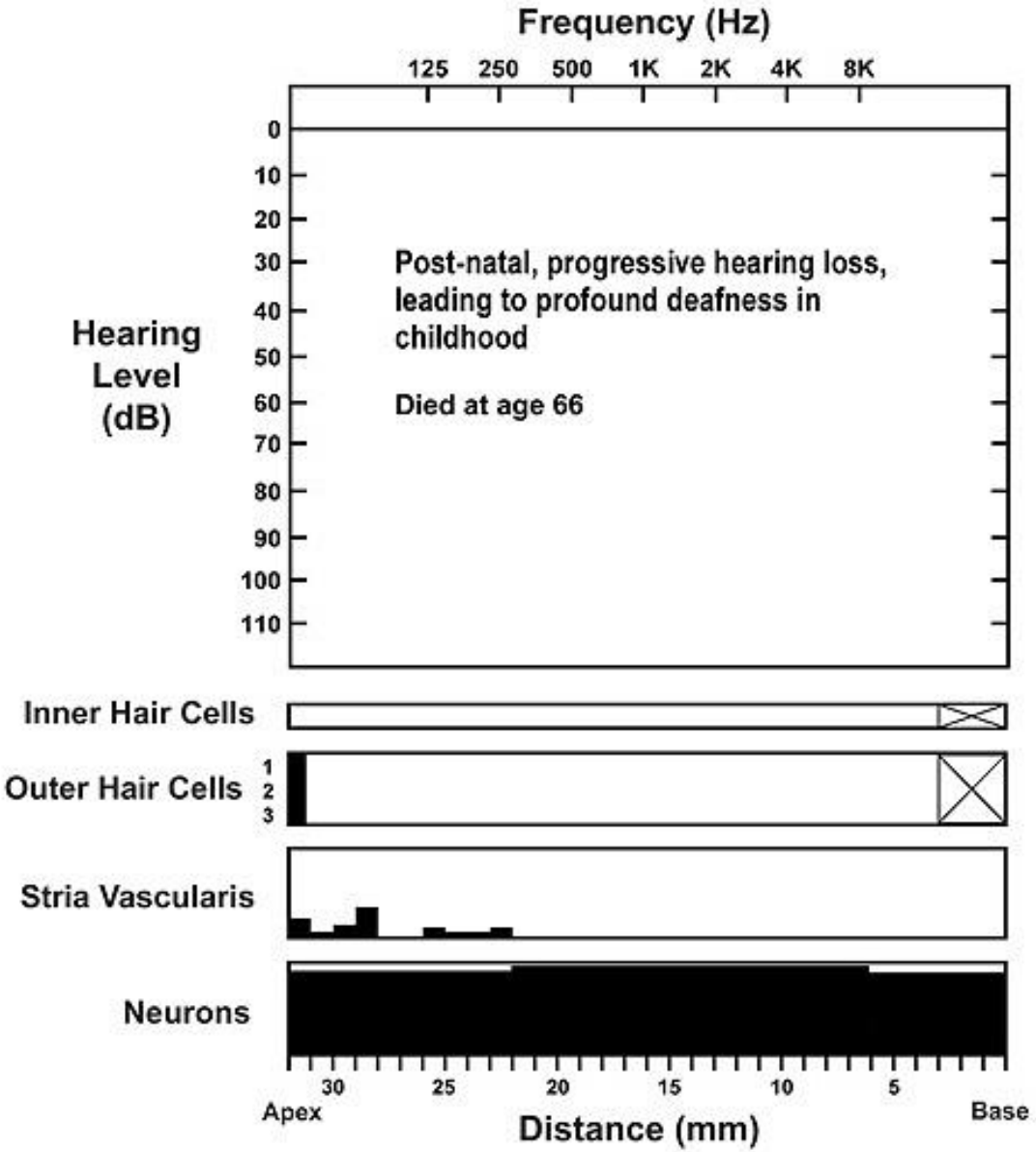
Comments: DFN-1, Mohr-Tranebjaerg Syndrome, Audio-cytocochleogram

Gender: Male

Age (yrs.): 67

Otologic Diagnosis:

1. Sensorineural hearing loss due to Mohr-Tranebjærg syndrome (DFN-1)
2. Degeneration, cochlear neurons, nearly total, secondary to #1
3. Degeneration, vestibular (Scarpa's) ganglion, severe, secondary to #1



Mohr-Tranebjaerg Syndrome, R-201

Title: Mohr-Tranebjaerg Syndrome, R-201

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mohr-Tranebjaerg Syndrome

TB Case Number: 920

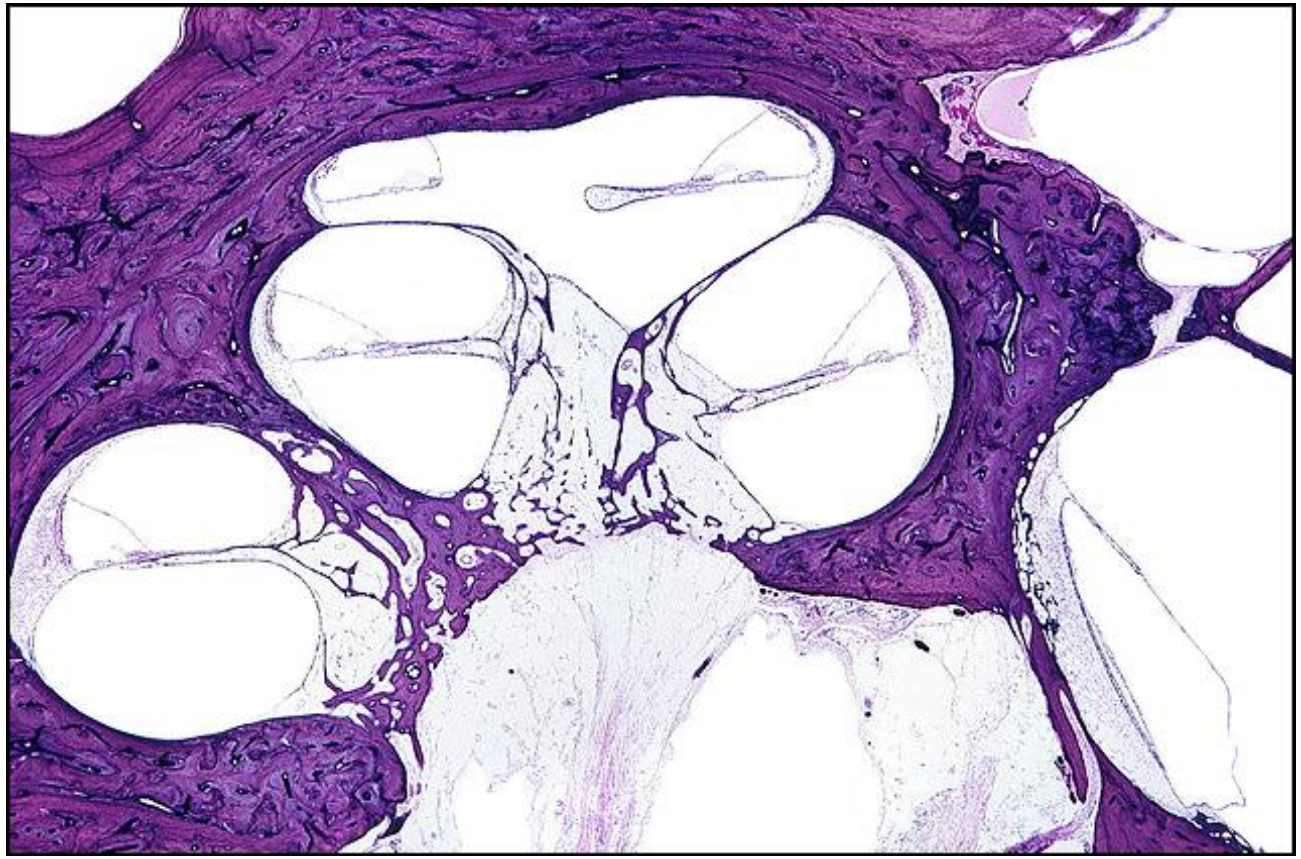
Comments: DFN-1, Mohr-Tranebjaerg Syndrome

Gender: Male

Age (yrs.): 67

Otologic Diagnosis:

1. Sensorineural hearing loss due to Mohr-Tranebjærg syndrome (DFN1)
2. Degeneration, cochlear neurons, nearly total, secondary to #1
3. Degeneration, vestibular (Scarpa's) ganglion, severe, secondary to #1



Mohr-Tranebjaerg Syndrome, R-201

Title: Mohr-Tranebjaerg Syndrome, R-201

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mohr-Tranebjaerg Syndrome

TB Case Number: 920

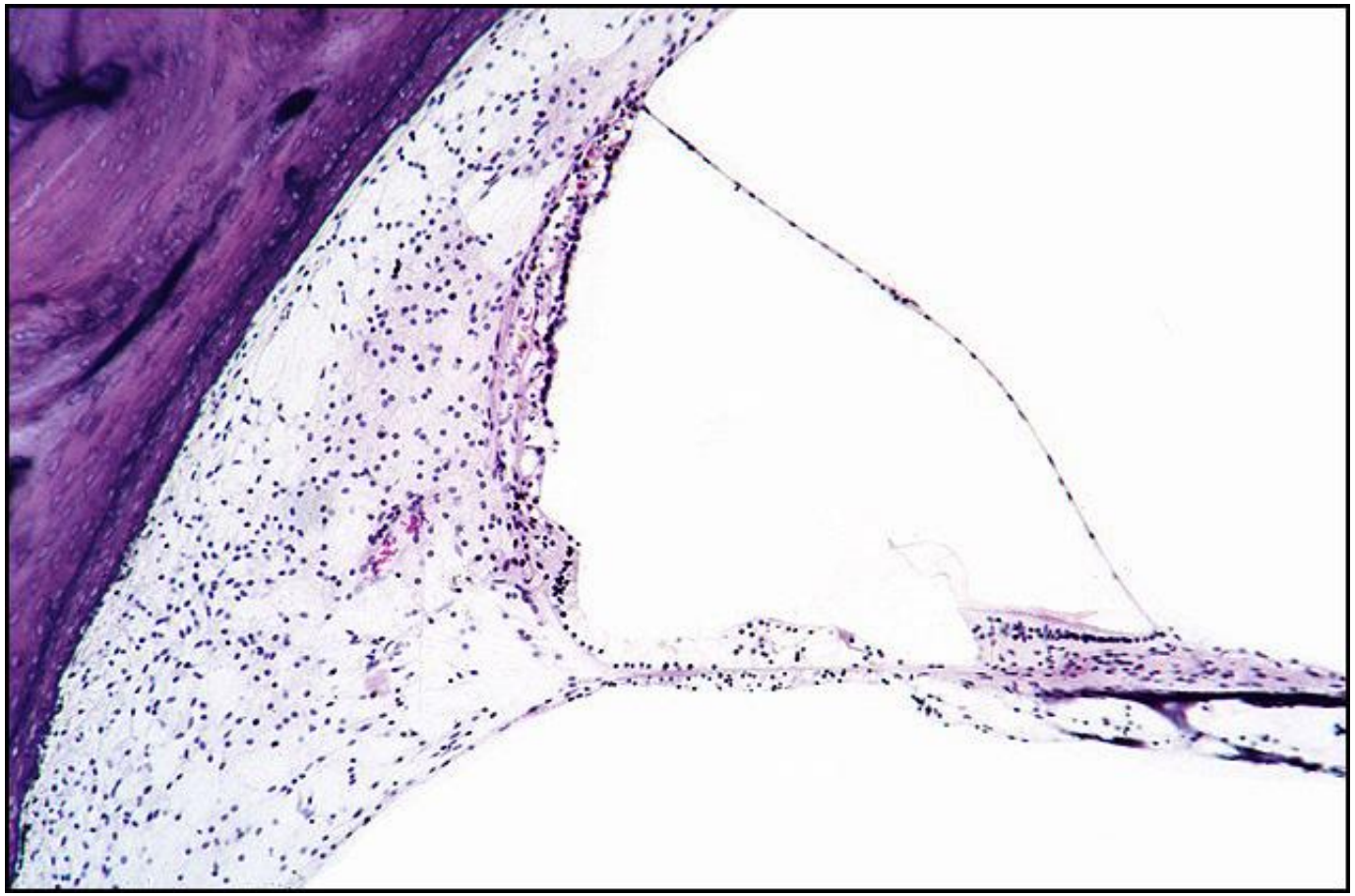
Comments: DFN-1, Mohr-Tranebjaerg Syndrome

Gender: Male

Age (yrs.): 67

Otologic Diagnosis:

1. Sensorineural hearing loss due to Mohr-Tranebjærg syndrome (DFN1)
2. Degeneration, cochlear neurons, nearly total, secondary to #1
3. Degeneration, vestibular (Scarpa's) ganglion, severe, secondary to #1



Mohr-Tranebjaerg Syndrome, R-11

Title: Mohr-Tranebjaerg Syndrome, R-11

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mohr-Tranebjaerg Syndrome

TB Case Number: 920

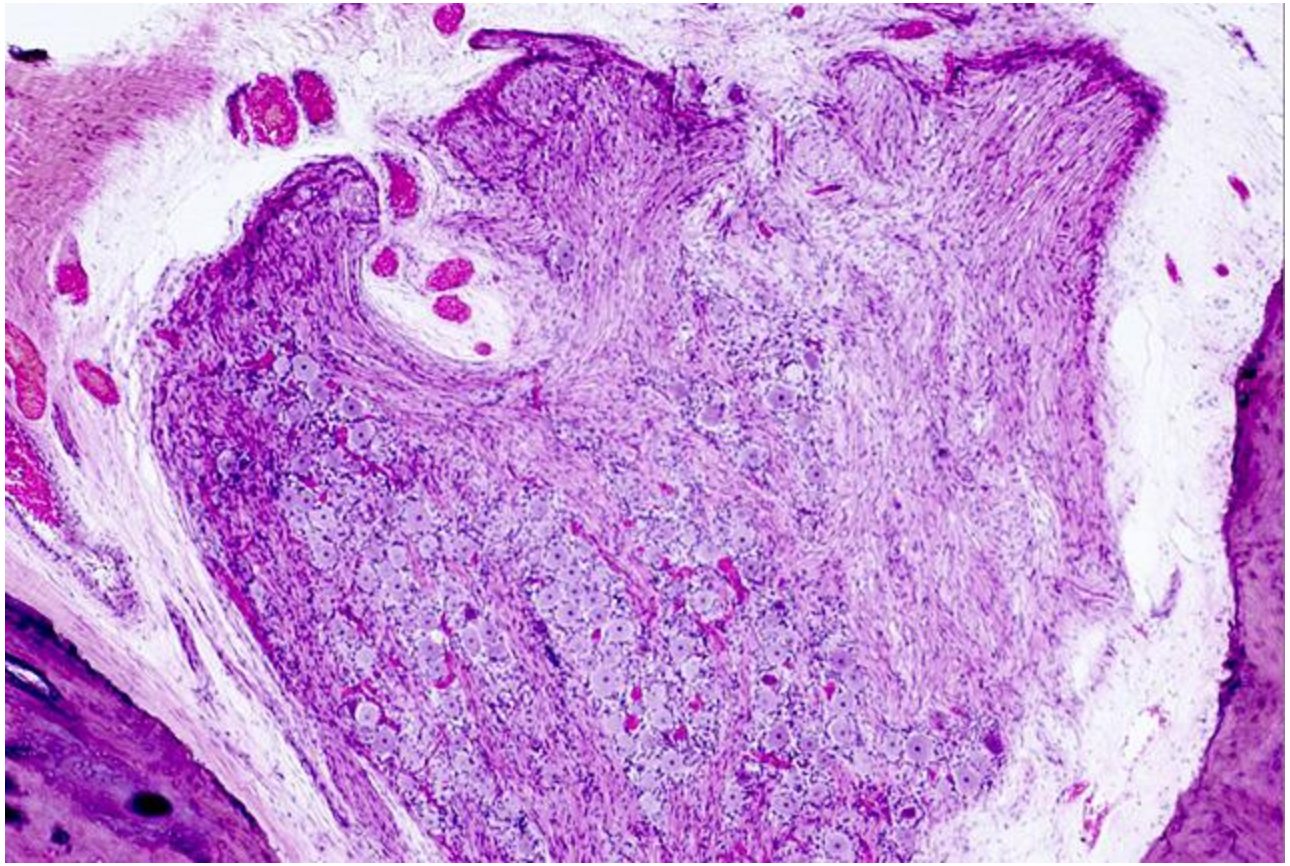
Comments: DFN-1, Mohr-Tranebjaerg Syndrome

Gender: Male

Age (yrs.): 67

Otologic Diagnosis:

1. Sensorineural hearing loss due to Mohr-Tranebjærg syndrome (DFN1)
2. Degeneration, cochlear neurons, nearly total, secondary to #1
3. Degeneration, vestibular (Scarpa's) ganglion, severe, secondary to #1



Mohr-Tranebjaerg Syndrome, R-201

Title: Mohr-Tranebjaerg Syndrome, R-201

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mohr-Tranebjaerg Syndrome

TB Case Number: 920

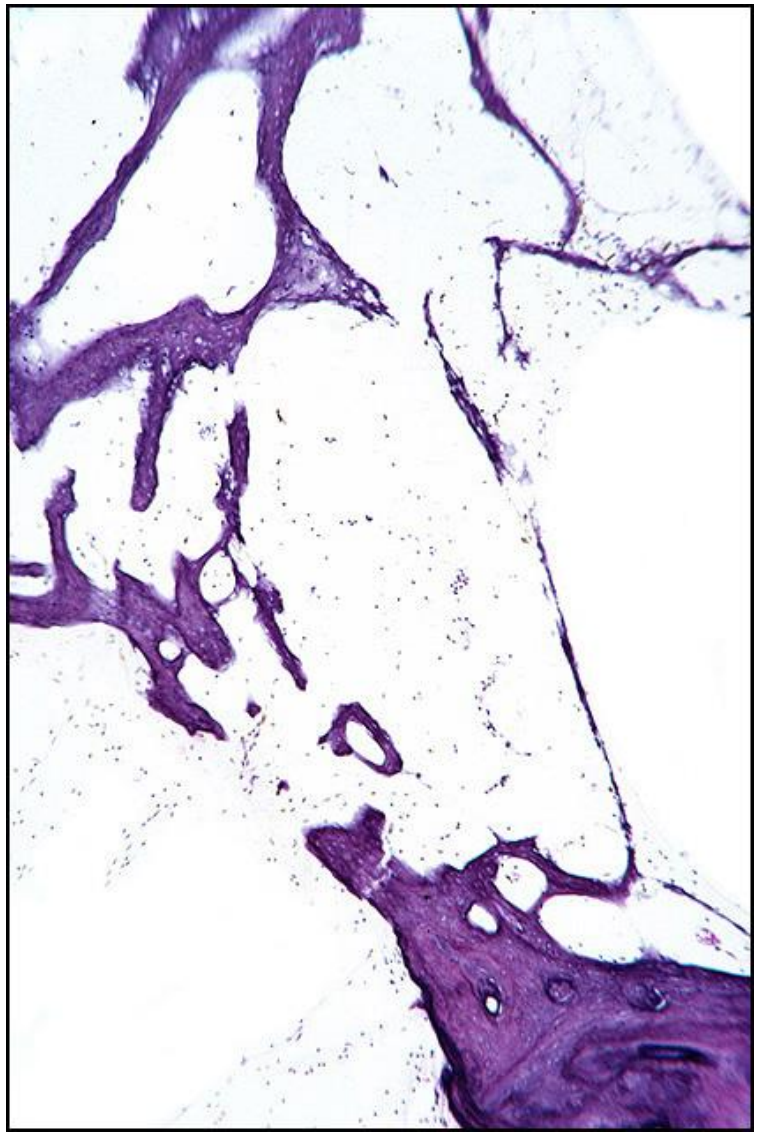
Comments: DFN-1, Mohr-Tranebjaerg Syndrome

Gender: Male

Age (yrs.): 67

Otologic Diagnosis:

1. Sensorineural hearing loss due to Mohr-Tranebjærg Syndrome (DFN1)
2. Degeneration, cochlear neurons, nearly total, secondary to #1
3. Degeneration, vestibular (Scarpa's) ganglion, severe, secondary to #1



MELAS, R-186

Title: MELAS, R-186

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss /Mitochondrial Hearing Loss - MELAS

TB Case Number: 919

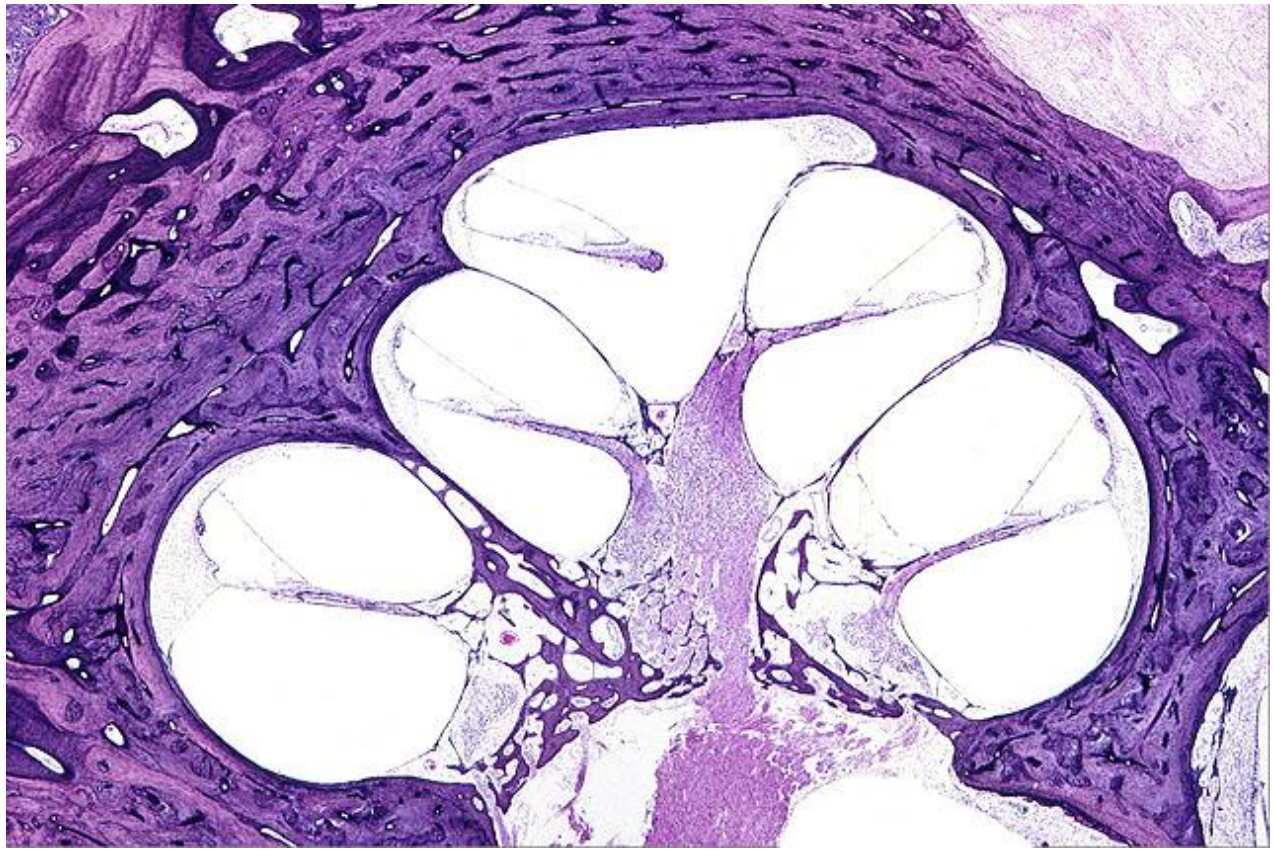
Comments: MELAS

Gender: Female

Age (yrs.): 23

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, resulting from MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes
2. Otitis media and mastoiditis, chronic, bilateral
3. Otitis media with effusion, bilateral
4. Stria vascularis, atrophy



MELAS

Title: MELAS

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss /Mitochondrial Hearing Loss - MELAS

TB Case Number: 919

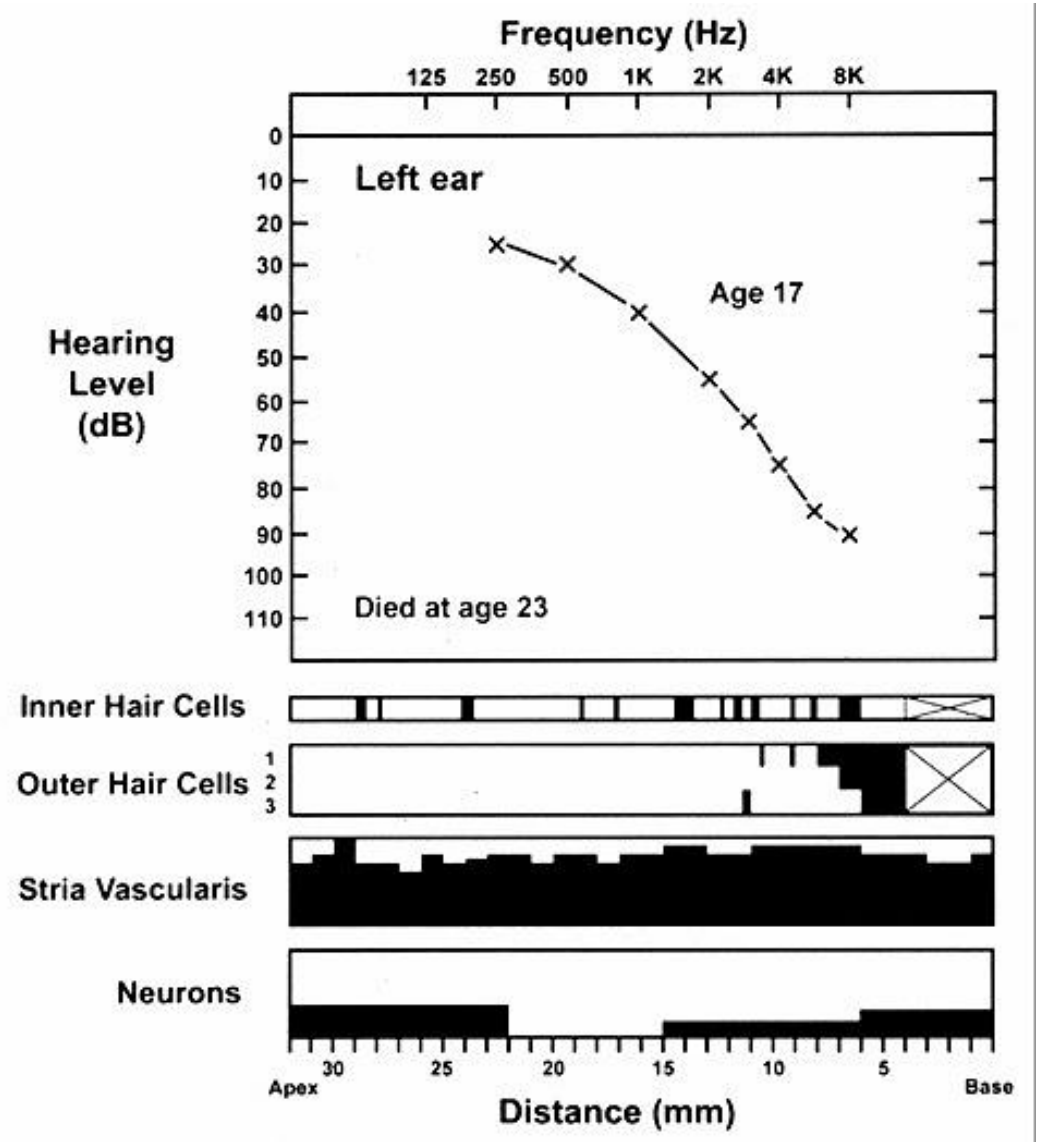
Comments: MELAS, Audio-cytocochleogram

Gender: Female

Age (yrs.): 23

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, resulting from MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes
2. Otitis media and mastoiditis, chronic, bilateral
3. Otitis media with effusion, bilateral
4. Stria vascularis, atrophy



MELAS

Title: MELAS

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss /Mitochondrial Hearing Loss - MELAS

TB Case Number: 919

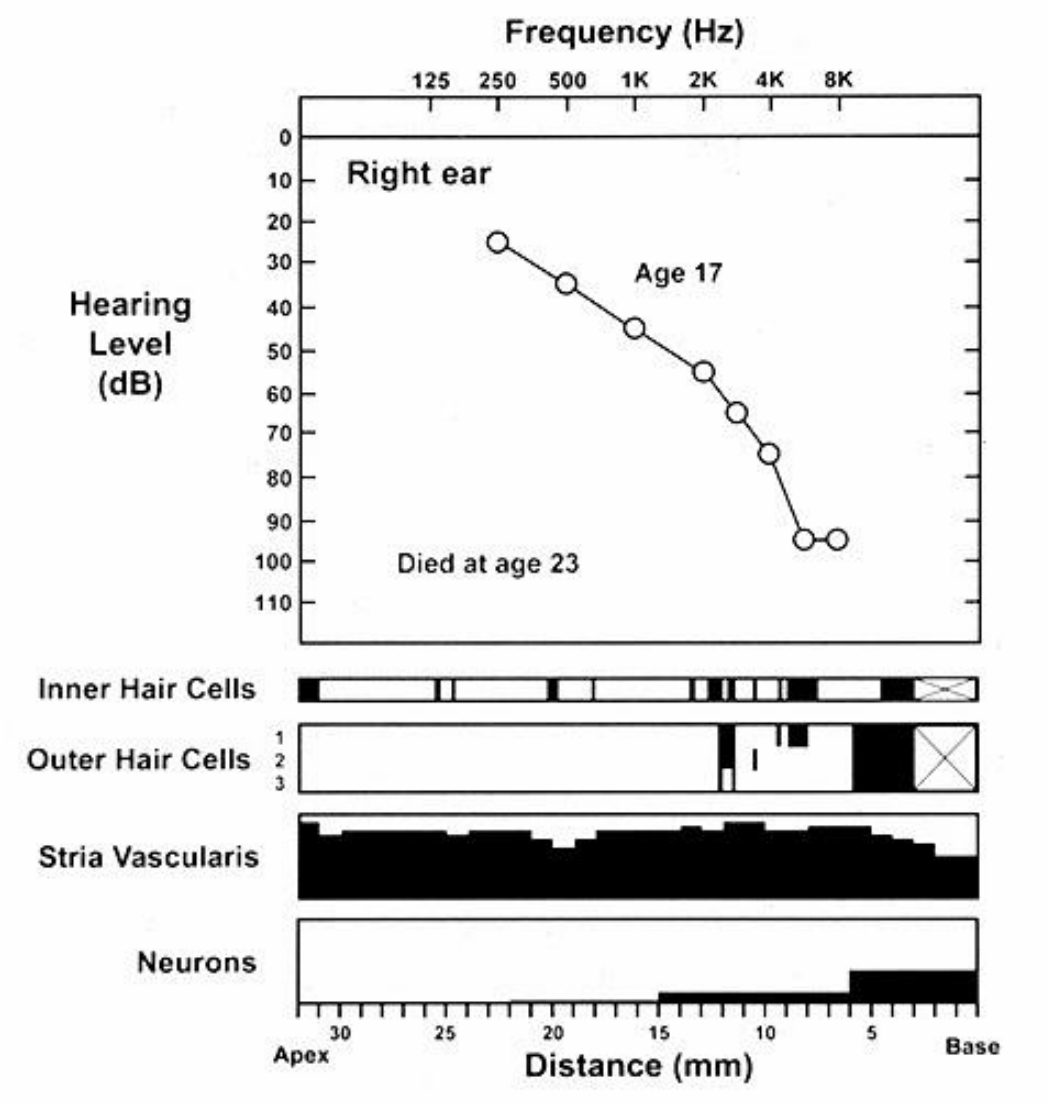
Comments: MELAS, Audio-cytocochleogram

Gender: Female

Age (yrs.): 23

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, resulting from MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes
2. Otitis media and mastoiditis, chronic, bilateral
3. Otitis media with effusion, bilateral
4. Stria vascularis, atrophy



MELAS, R-281

Title: MELAS, R-281

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss /Mitochondrial Hearing Loss - MELAS

TB Case Number: 919

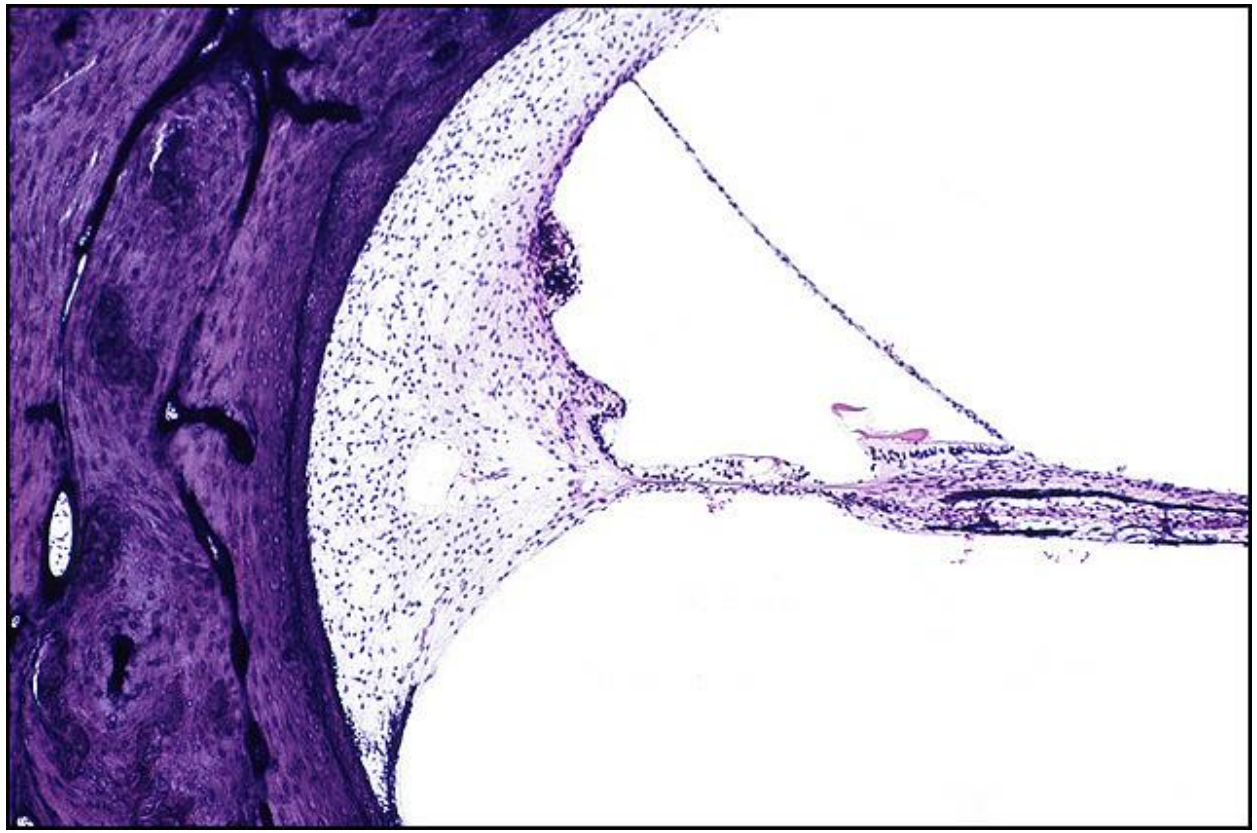
Comments: MELAS

Gender: Female

Age (yrs.): 23

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, resulting from MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes
2. Otitis media and mastoiditis, chronic, bilateral
3. Otitis media with effusion, bilateral
4. Stria vascularis, atrophy



MELAS, R-186

Title: MELAS, R-186

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss /Mitochondrial Hearing Loss - MELAS

TB Case Number: 919

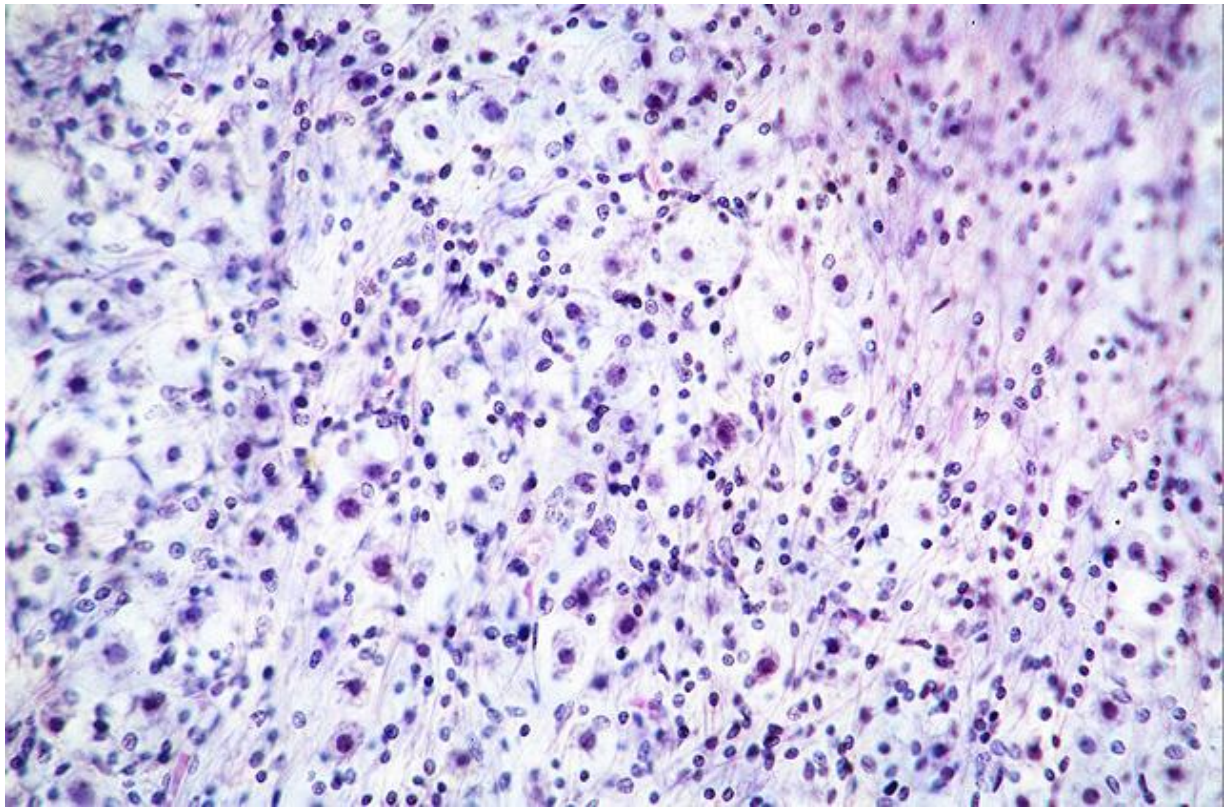
Comments: MELAS

Gender: Female

Age (yrs.): 23

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, resulting from MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes
2. Otitis media and mastoiditis, chronic, bilateral
3. Otitis media with effusion, bilateral
4. Stria vascularis, atrophy



MELAS, R-41

Title: MELAS, R-41

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mitochondrial Hearing Loss - MELAS

TB Case Number: 919

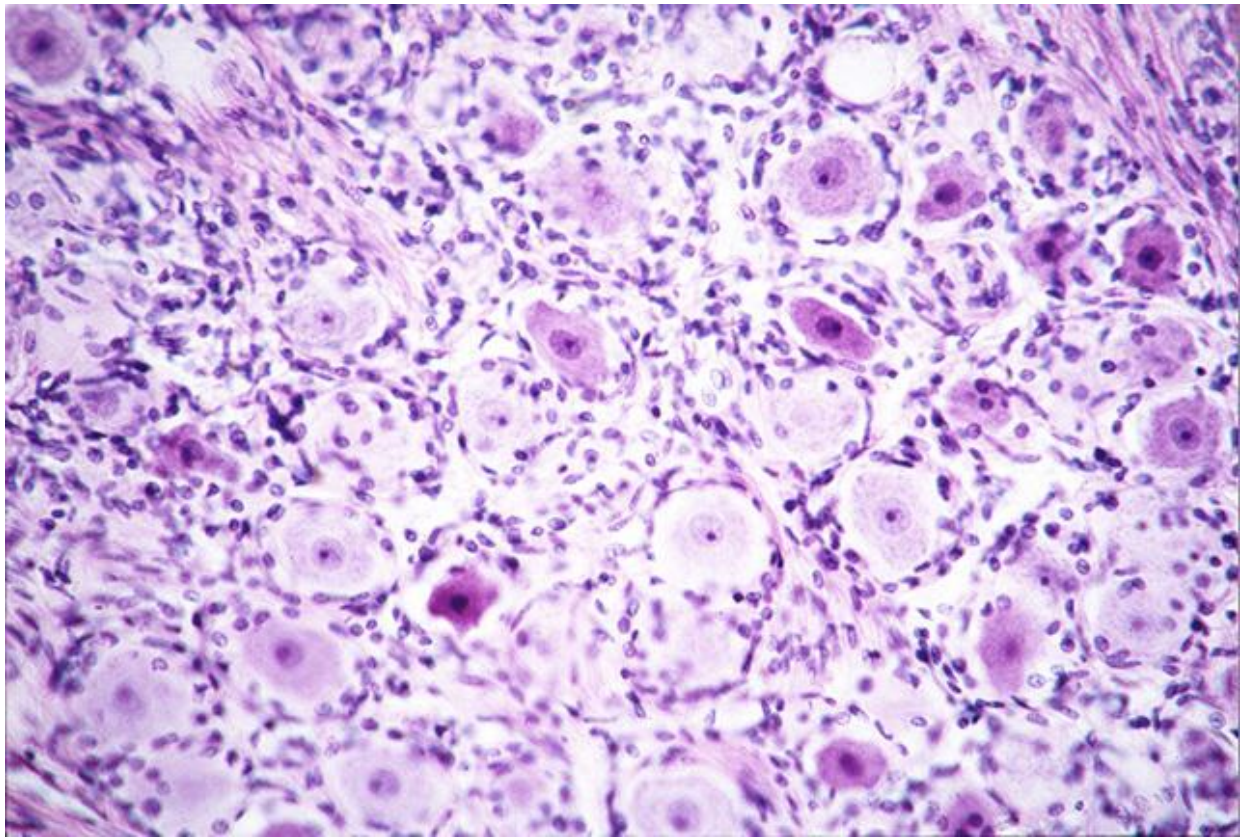
Comments: MELAS

Gender: Female

Age (yrs.): 23

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, resulting from MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes
2. Otitis media and mastoiditis, chronic, bilateral
3. Otitis media with effusion, bilateral
4. Stria vascularis, atrophy



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 948

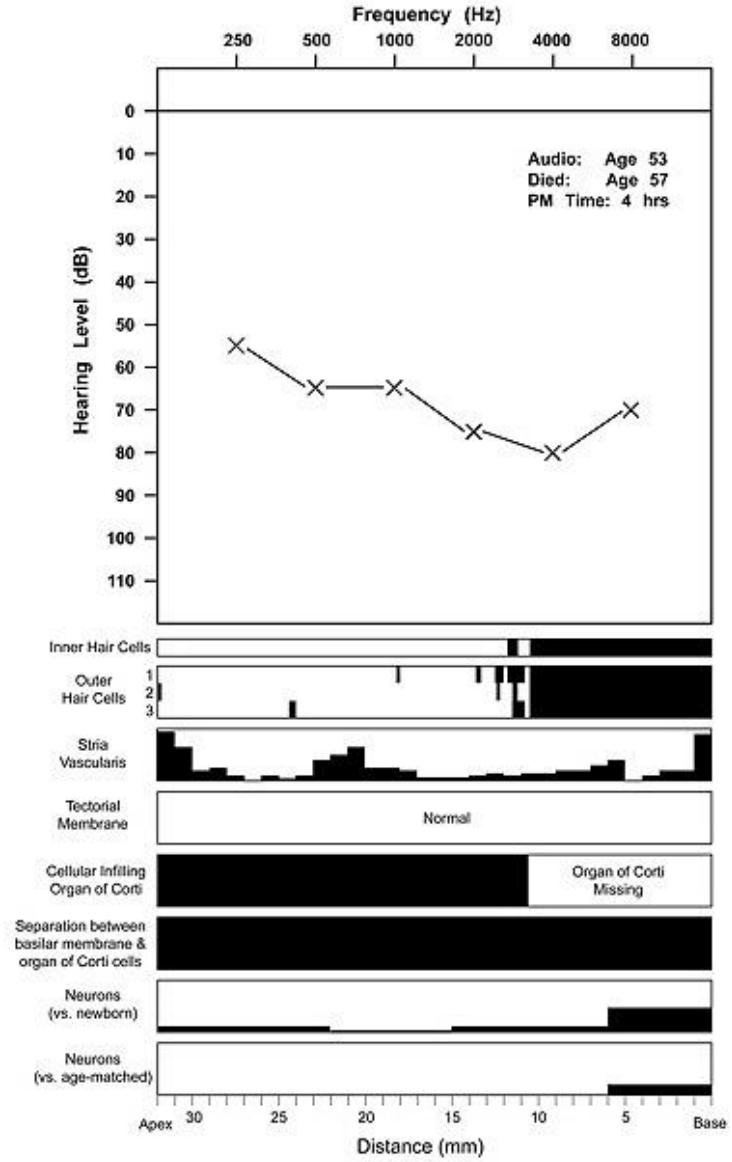
Comments: Alport syndrome Audio-cytocochleogram

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 948

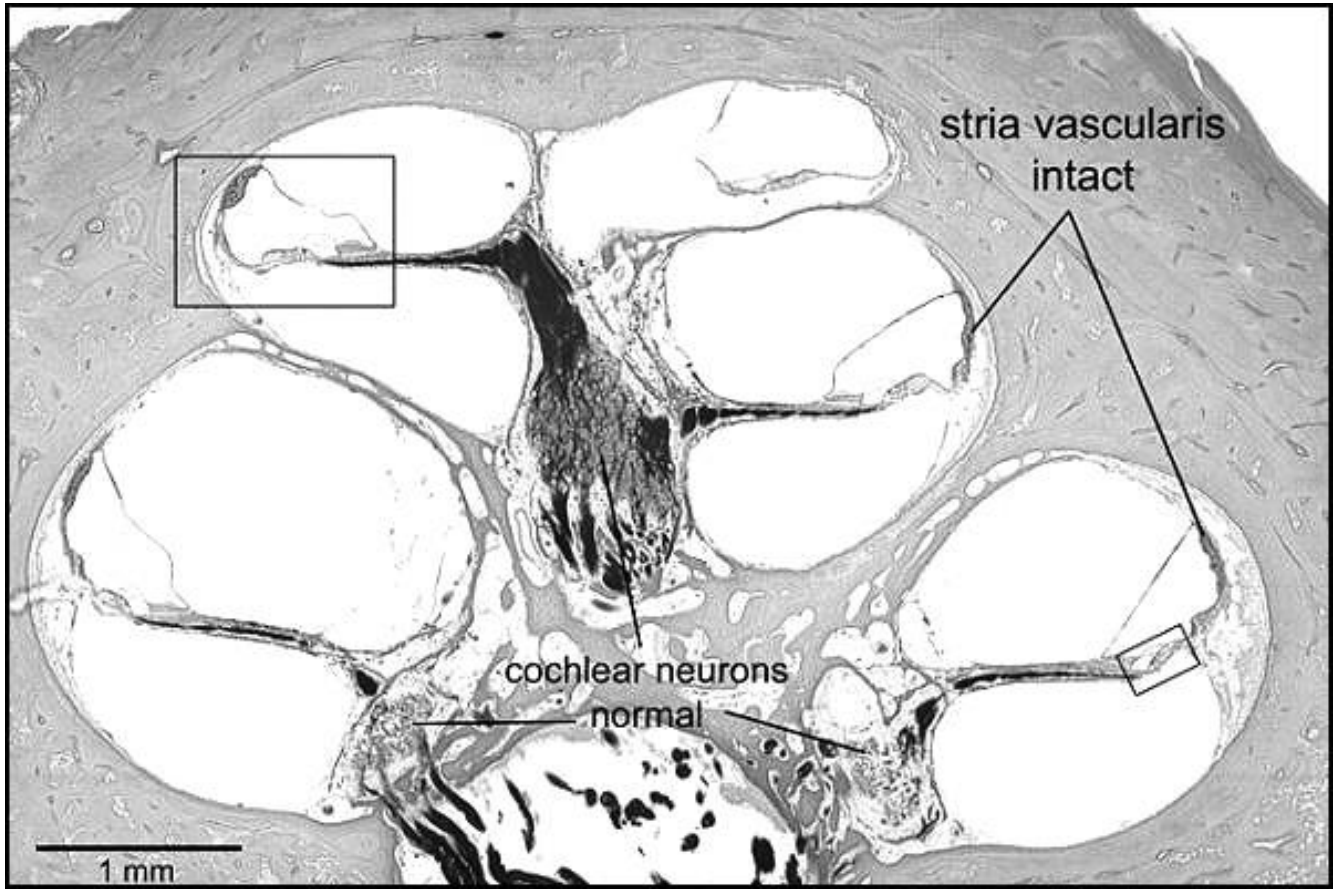
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 948

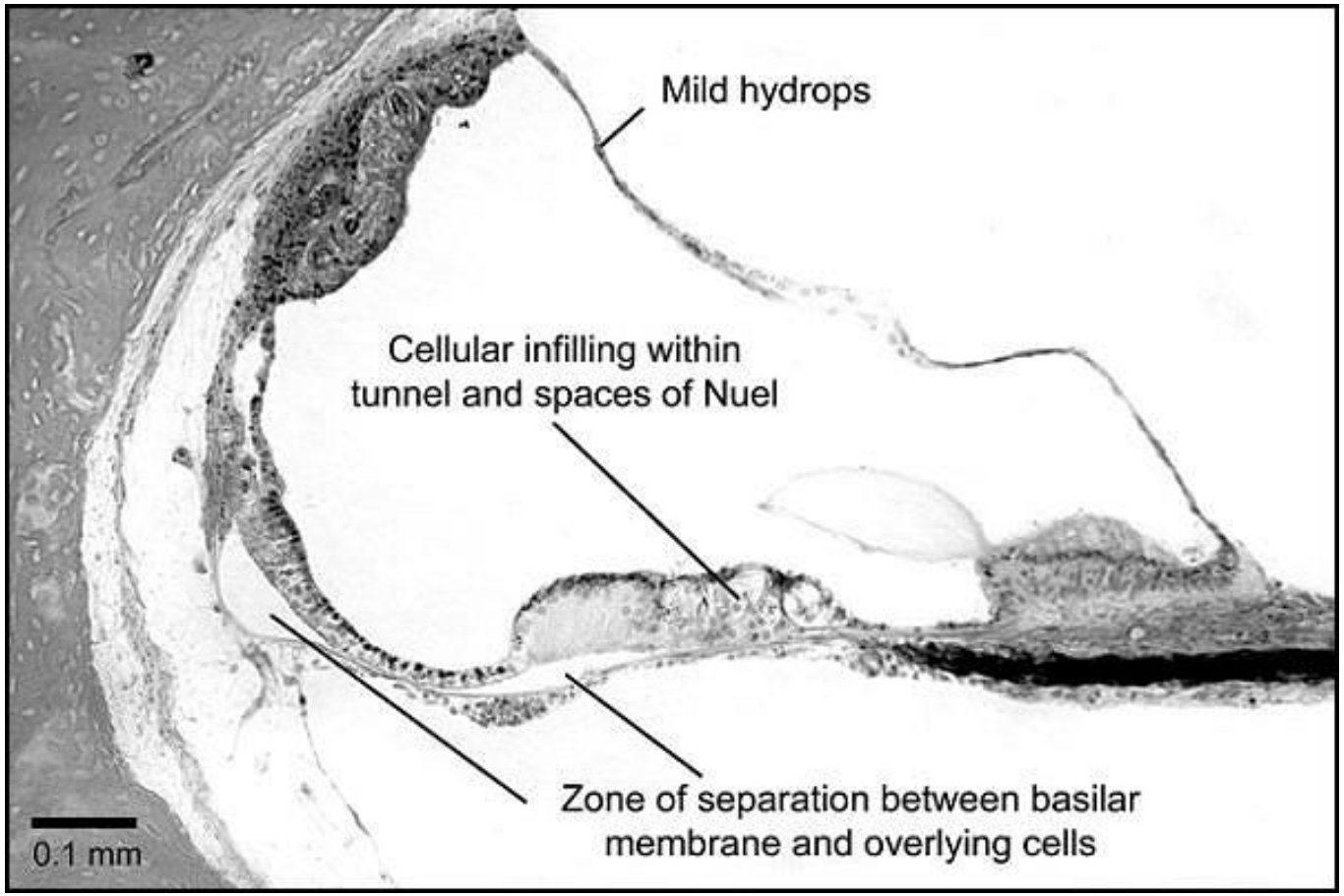
Comments: Alport syndrome

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 948

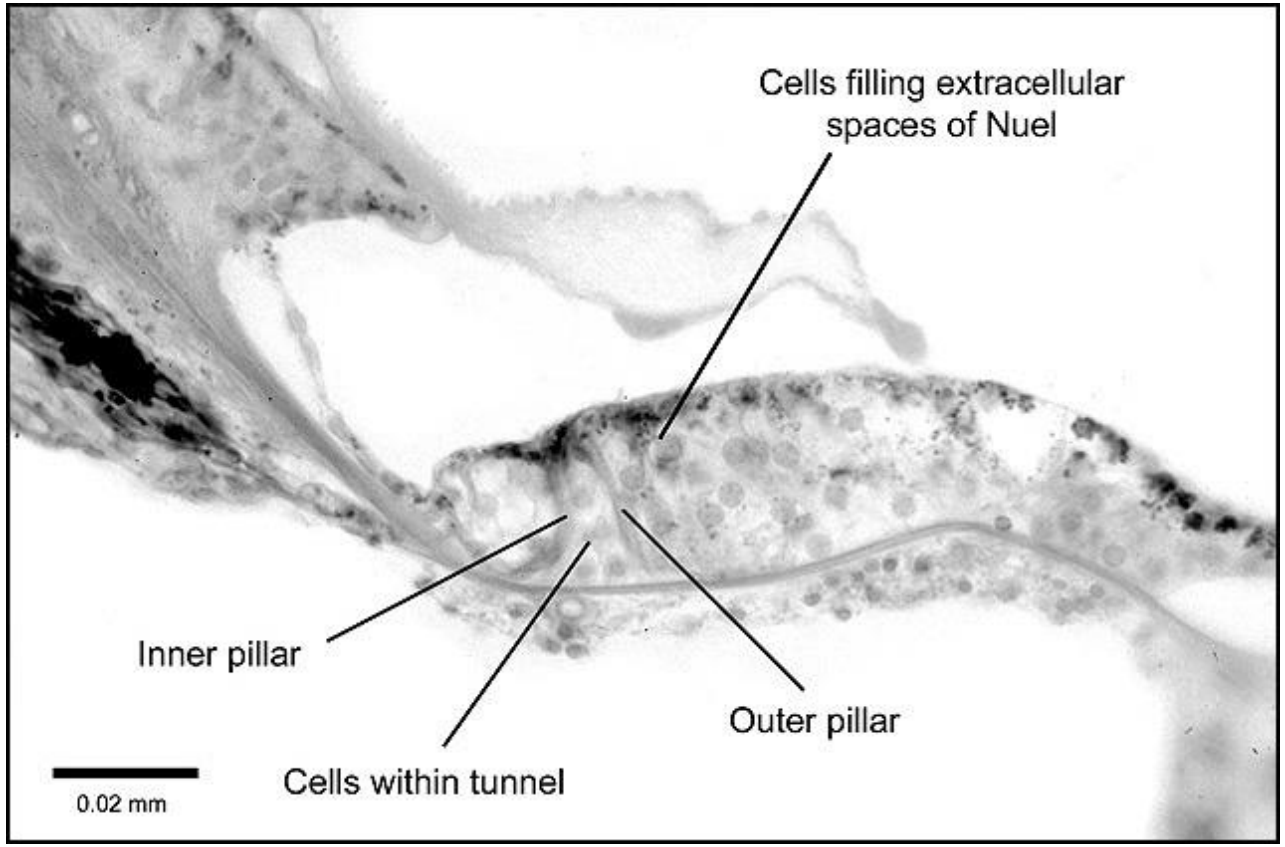
Comments: Alport syndrome

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss /Alport Syndrome

TB Case Number: 948

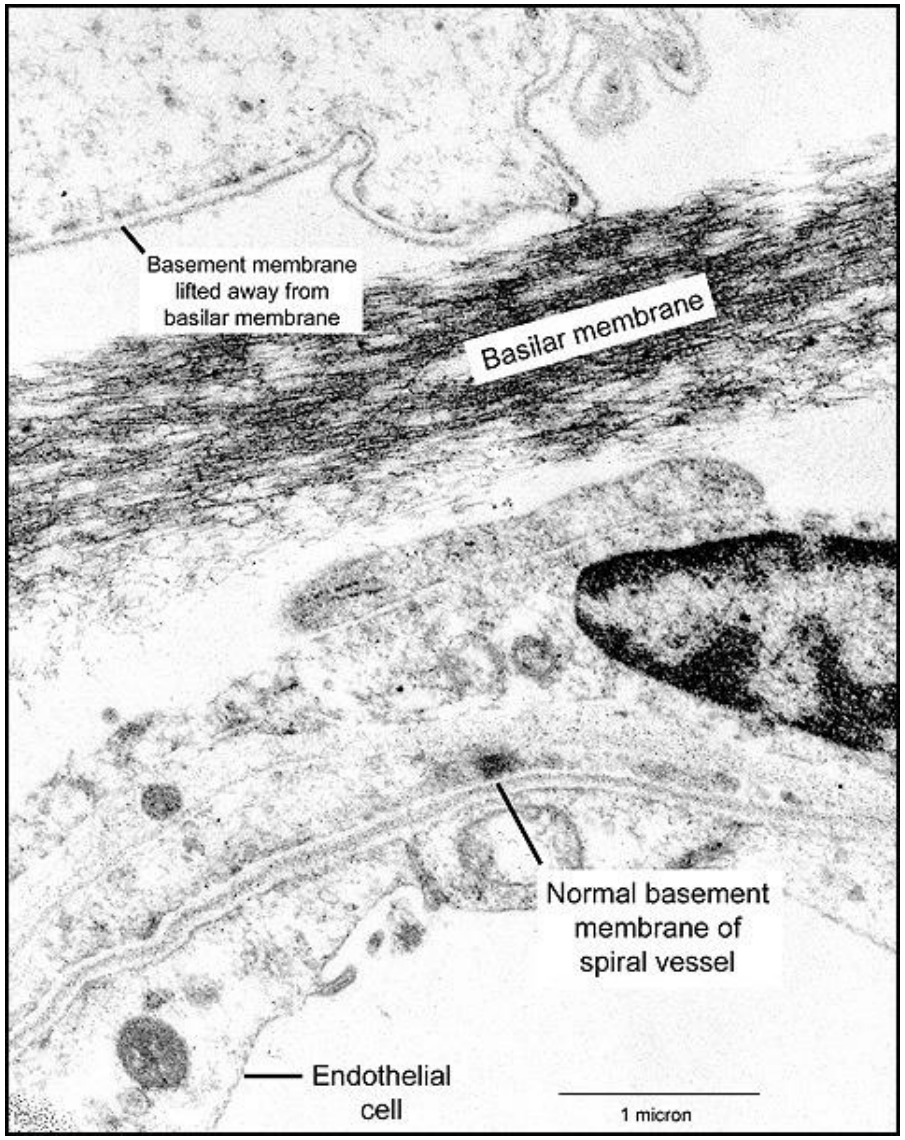
Comments: Alport syndrome

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 948

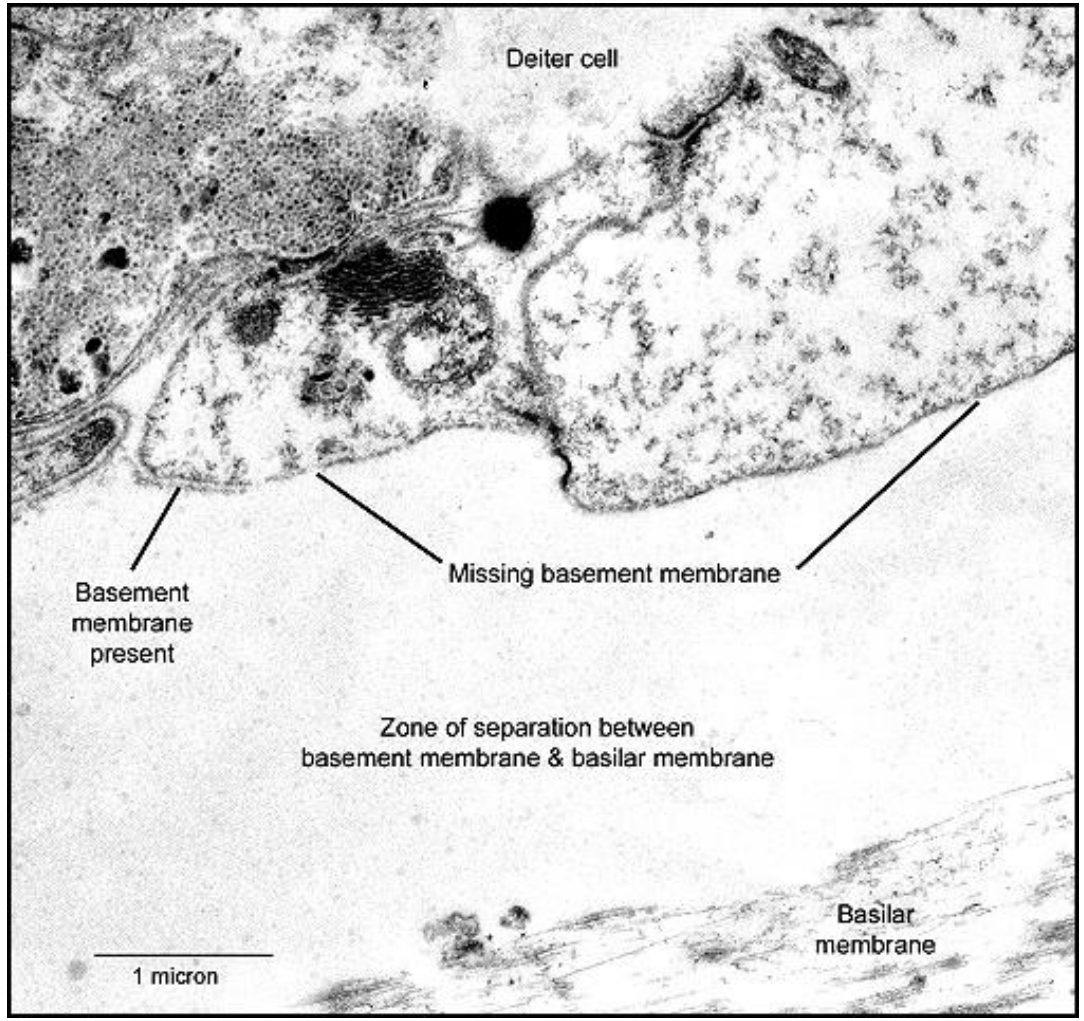
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 948

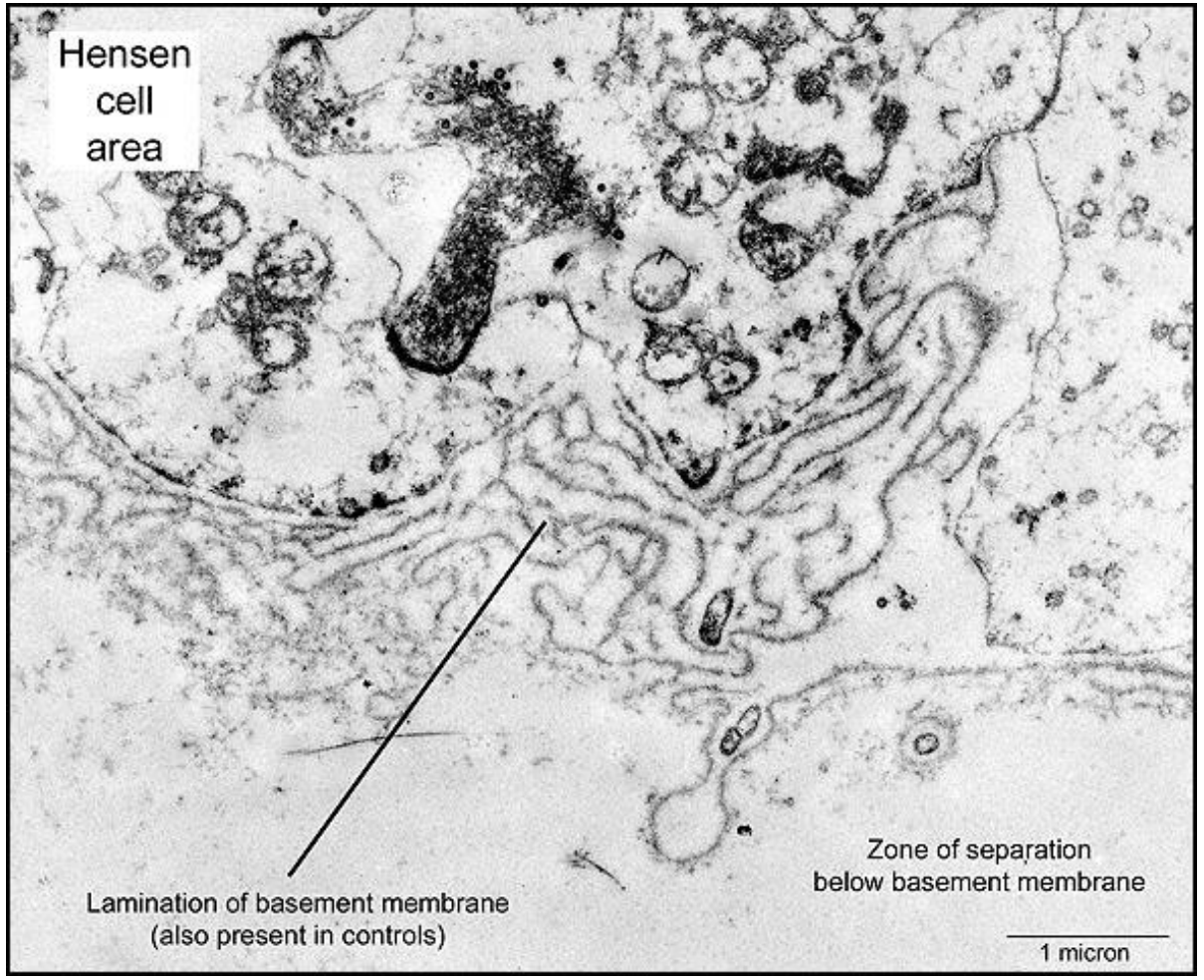
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss /Alport Syndrome

TB Case Number: 948

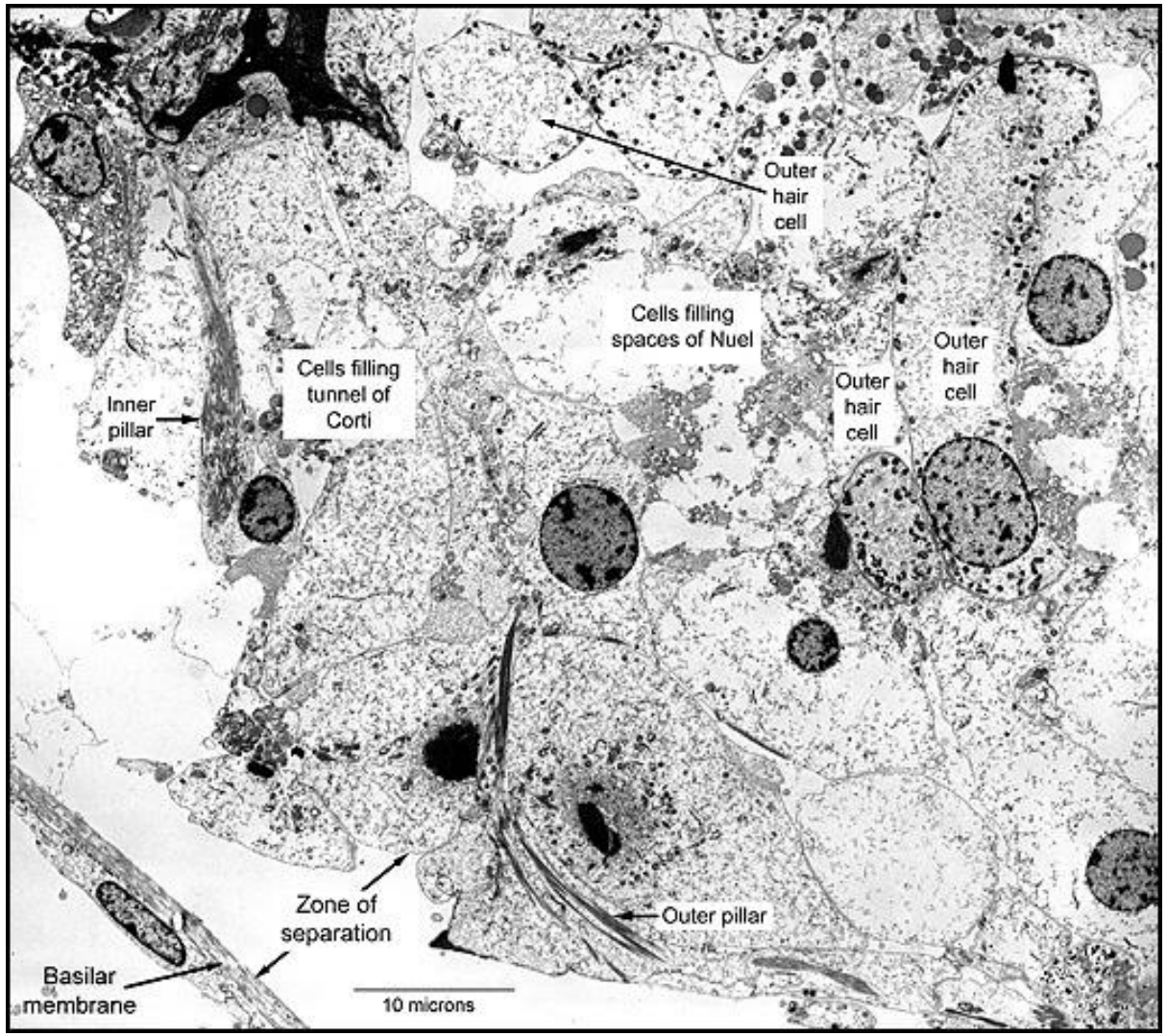
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 948

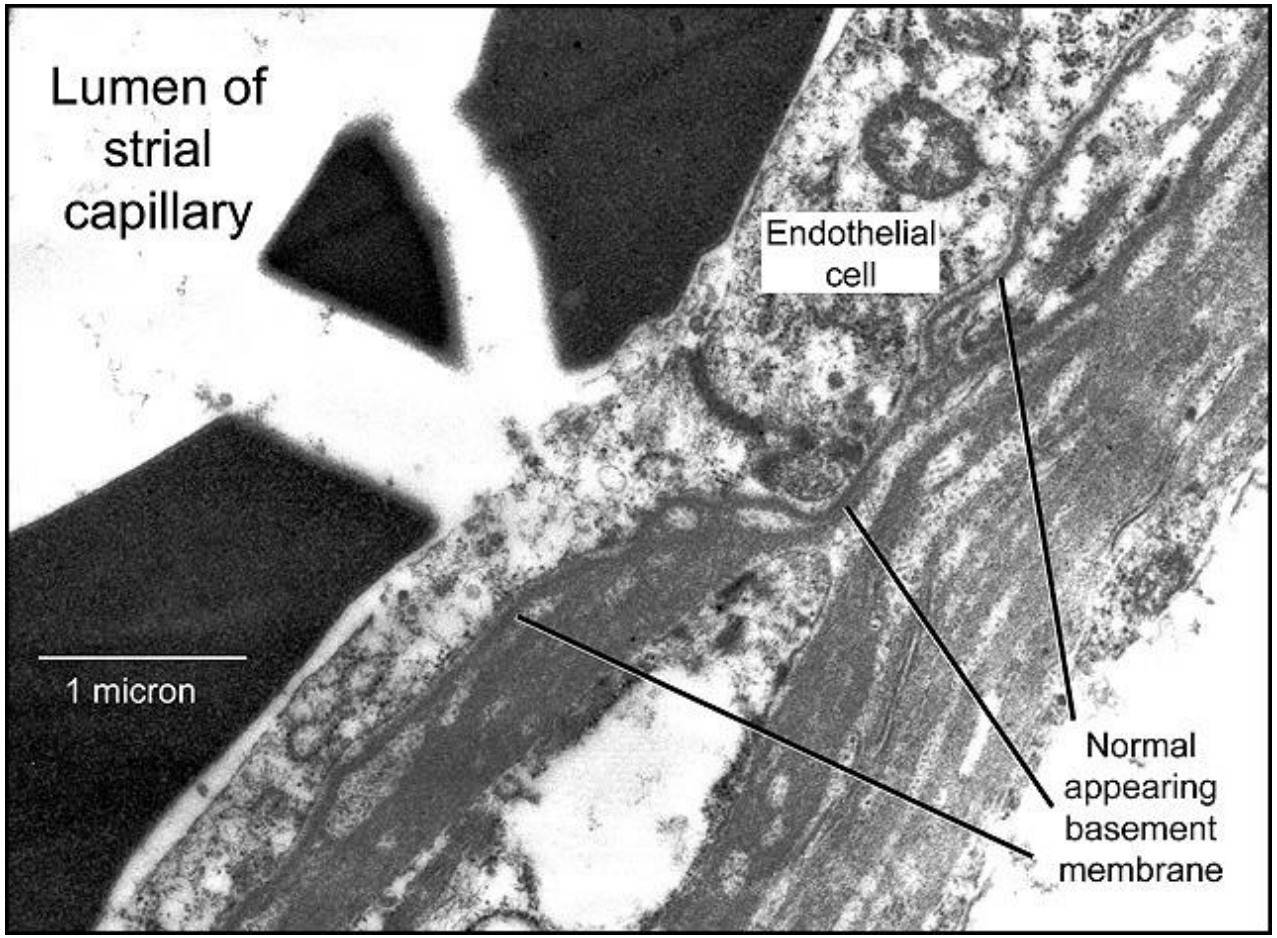
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 58

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, associated with X-linked Alport syndrome due to COL4A5 mutation on Xq22
2. Endolymphatic hydrops, bilateral (mild on left, moderate-to-severe on right)
3. Organ of Corti, hair cell loss, bilateral



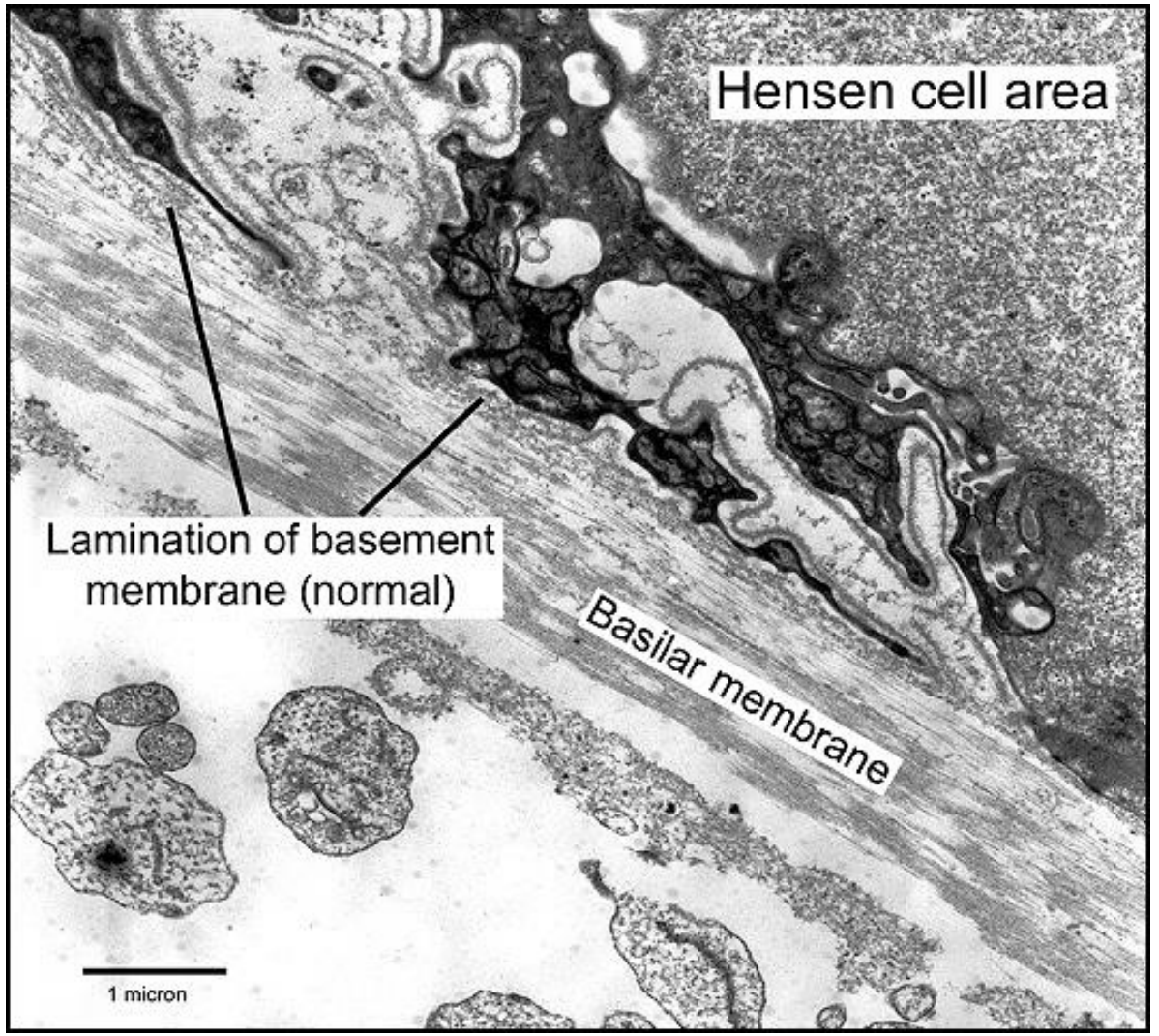
Alport Syndrome, Control

Title: Alport Syndrome, Control

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Alport Syndrome

Comments: Control-H18, Male-53



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 954

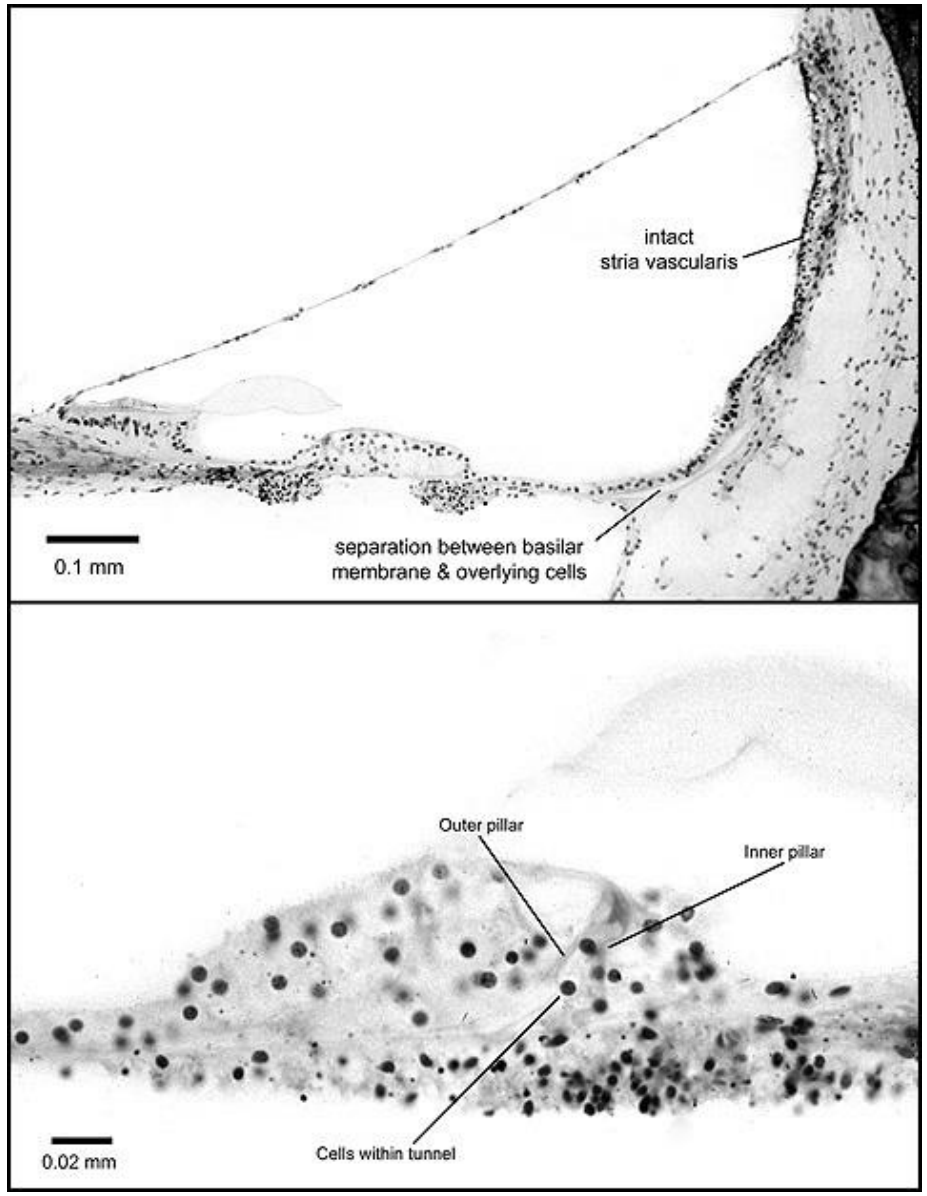
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 60

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, with hemorrhagic nephritis (Alport syndrome)
2. Zone of separation between basilar membrane and overlying cells (presumed basement membrane defect), all cochlear turns, bilateral
3. Organ of Corti, dysmorphogenesis



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 801

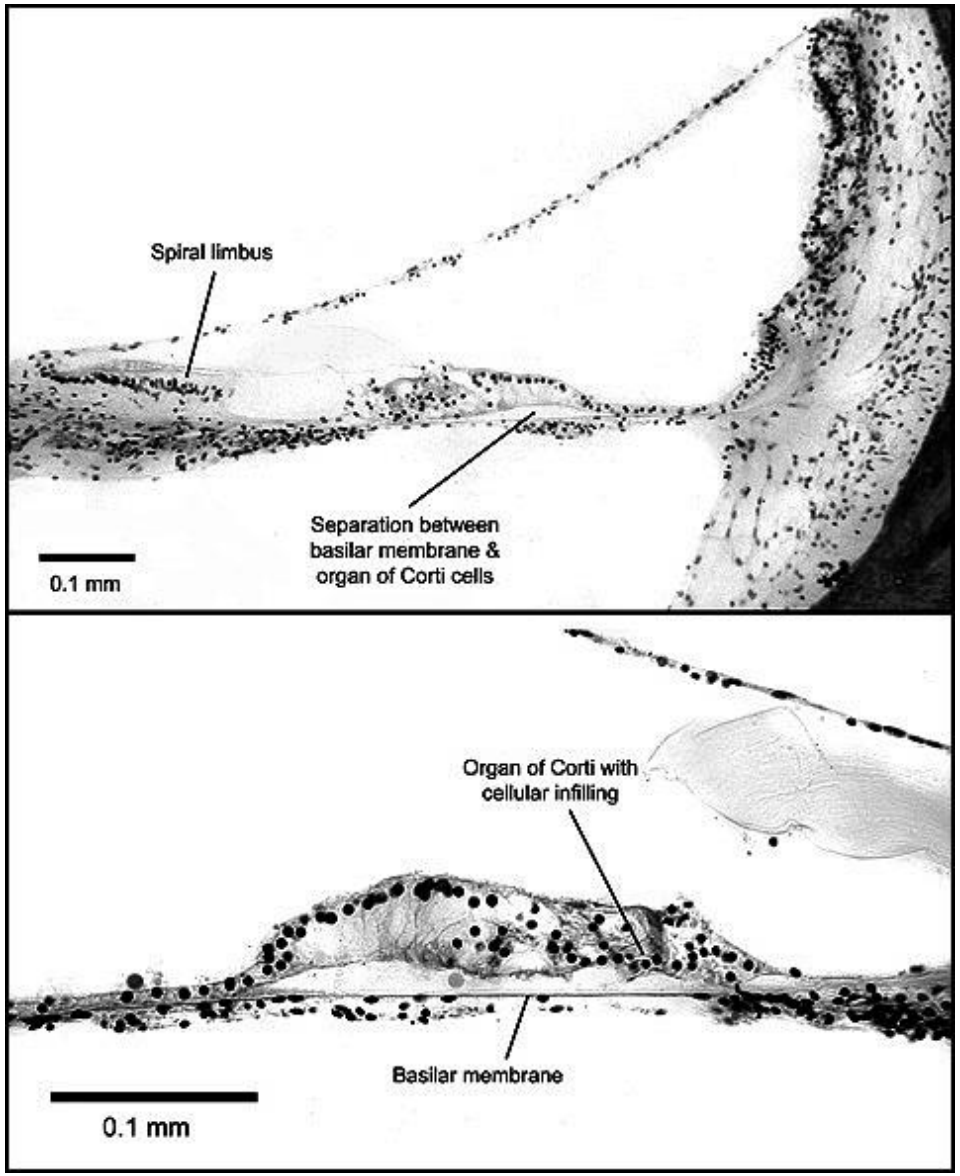
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 50

Otologic Diagnosis:

1. Alport's syndrome, with hearing loss by history (no audiogram) and renal failure
2. Lymphomatoid granulomatosis as manifested by perivascular infiltrates of the vessels of the internal auditory canal and infiltrates in the nerve trunks.



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 727

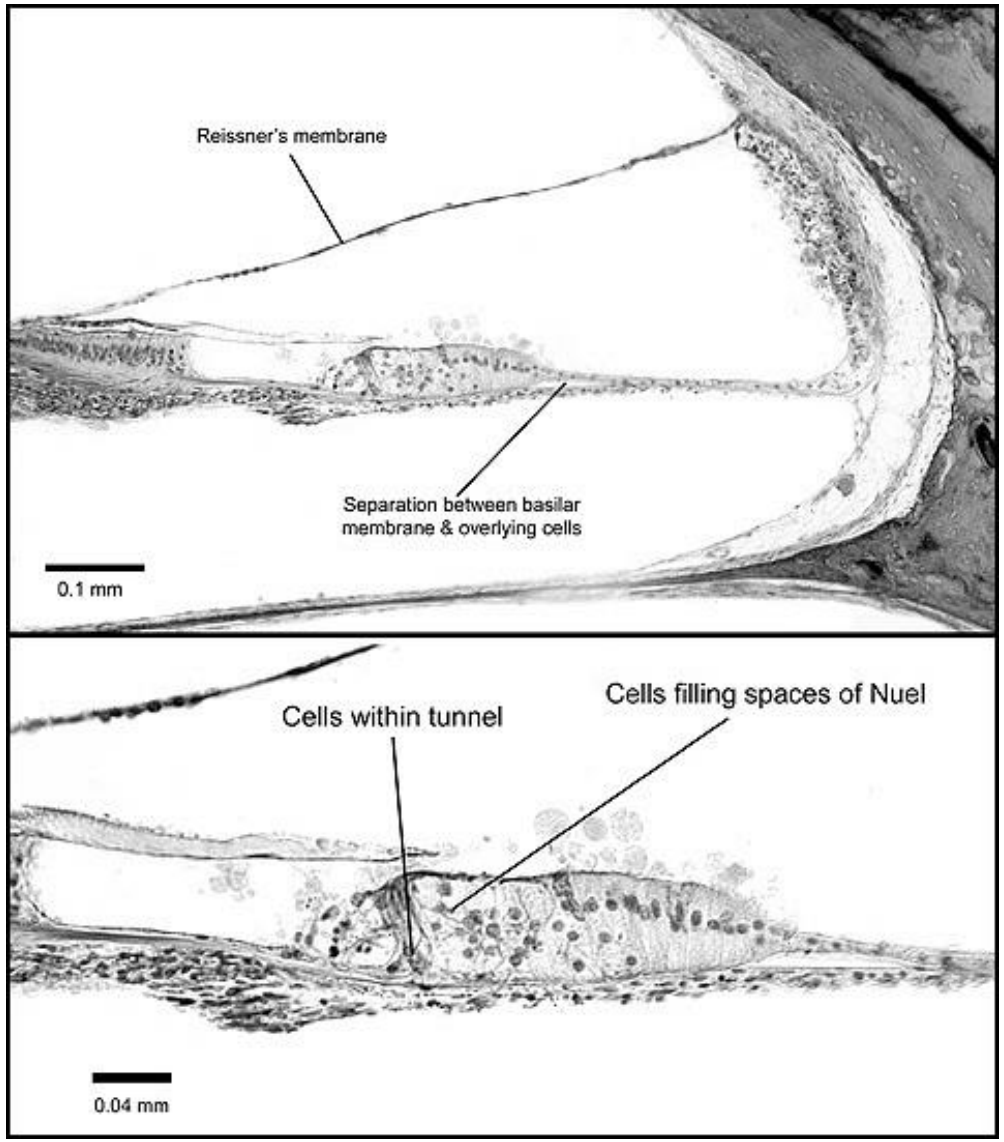
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 20

Otologic Diagnosis:

1. Alport's syndrome
2. Otosclerosis, without stapes fixation, bilateral



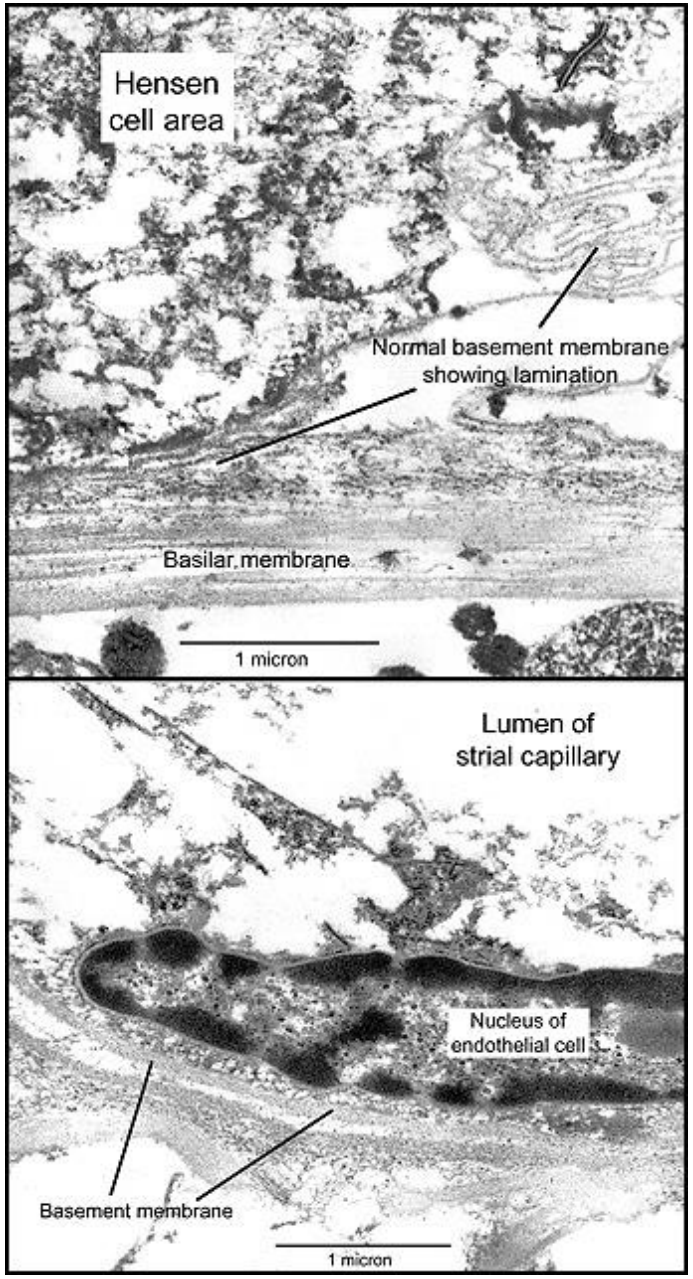
Alport Syndrome, Control

Title: Alport Syndrome, Control

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Alport Syndrome

Comments: Control



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 954

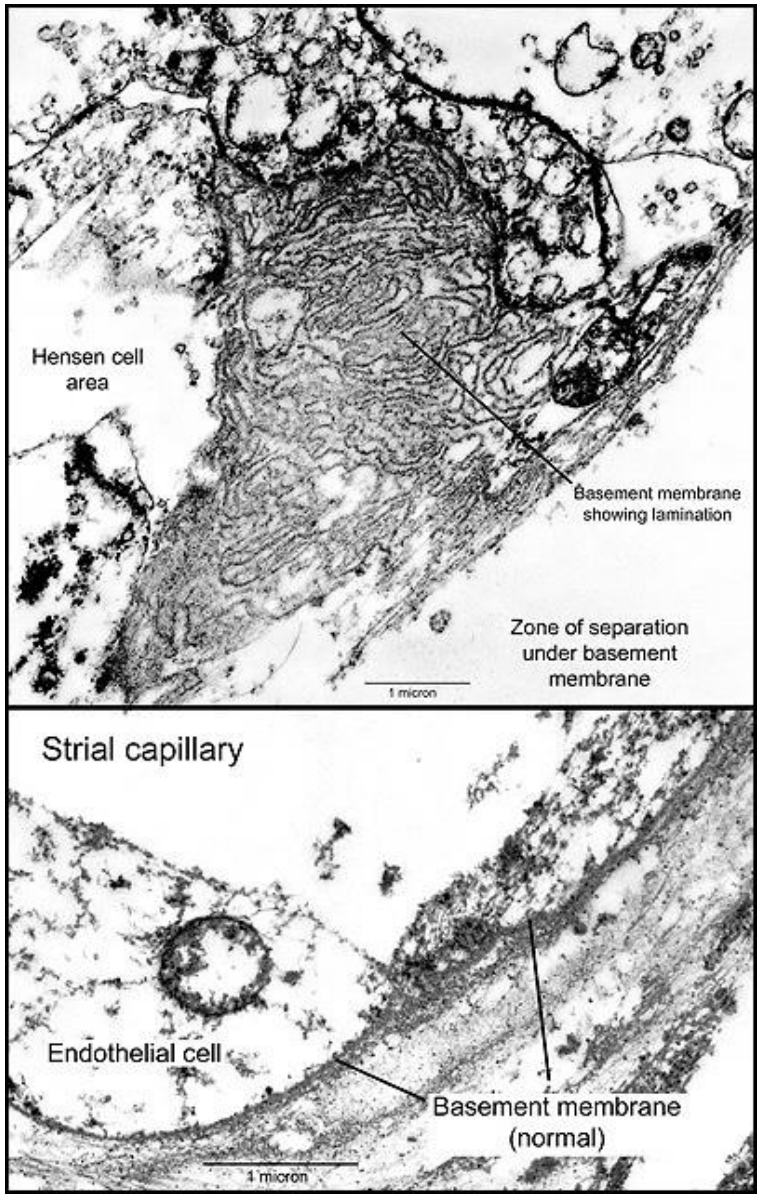
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 60

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, with hemorrhagic nephritis (Alport syndrome)
2. Zone of separation between basilar membrane and overlying cells (presumed basement membrane defect), all cochlear turns, bilateral
3. Organ of Corti, dysmorphogenesis



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

TB Case Number: 504

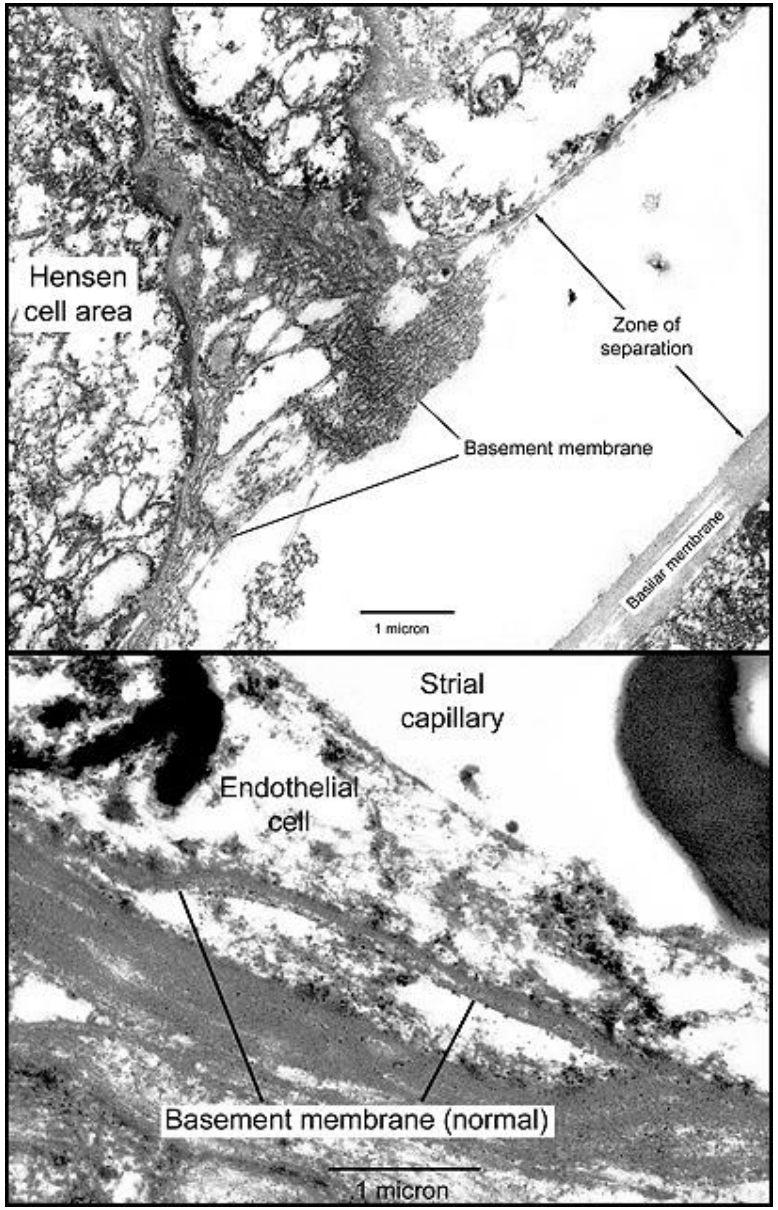
Comments: Alport Syndrome

Gender: Male

Age (yrs.): 49

Otologic Diagnosis:

1. Alport's syndrome, atypical, late onset of hearing loss
2. Sensorineural hearing loss, associated with Alport's syndrome, bilateral
3. Organ of Corti, degeneration, severe, basal turn, bilateral
4. Otitis media with effusion, tympanomastoid compartment



Alport Syndrome, L-201.3

Title: Alport syndrome, L-201.3

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

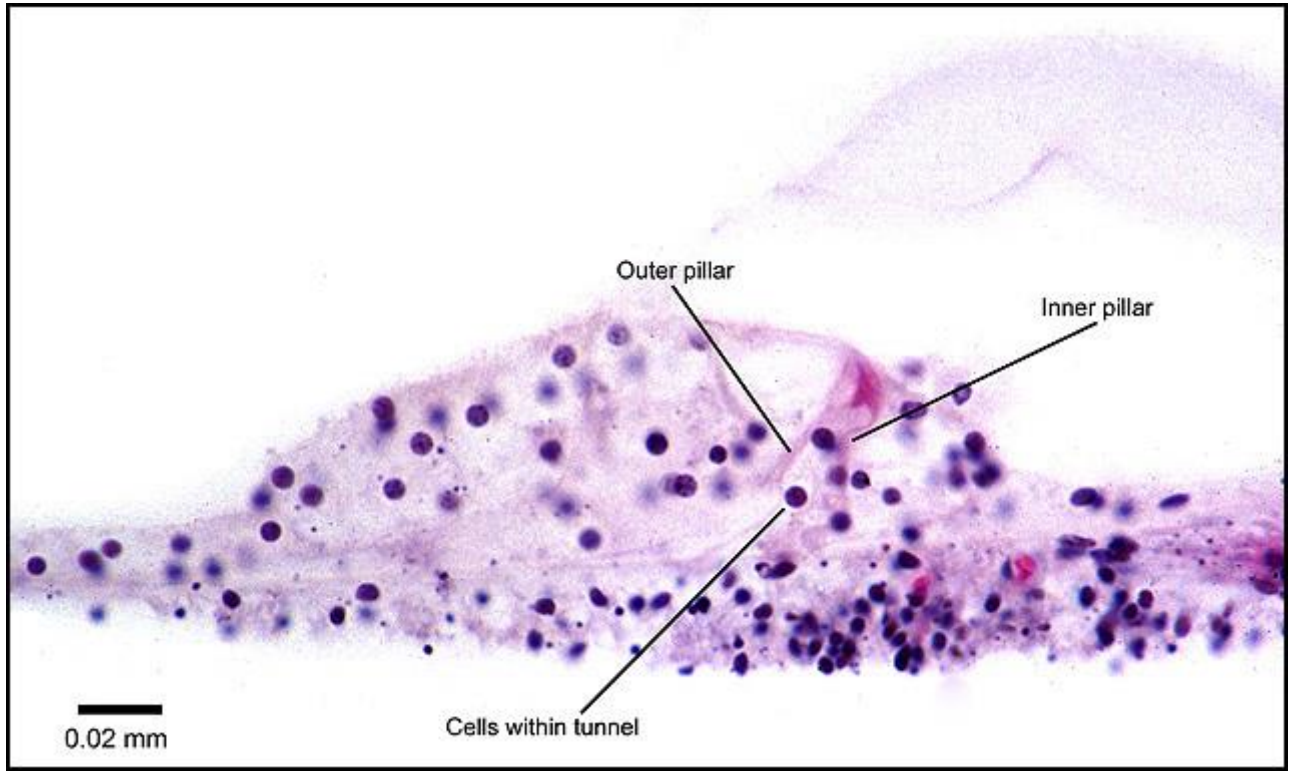
TB Case Number: 954

Gender: Male

Age (yrs.): 60

Otologic Diagnosis:

1. Sensorineural hearing loss, bilateral, with hemorrhagic nephritis (? Alport syndrome)
2. Zone of separation between basilar membrane and overlying cells (presumed basement membrane defect), all cochlear turns, bilateral
3. Organ of Corti, dysmorphogenesis



Alport Syndrome

Title: Alport Syndrome

Chapter: Developmental Defects

Chapter section: Syndromic Hearing Loss / Alport Syndrome

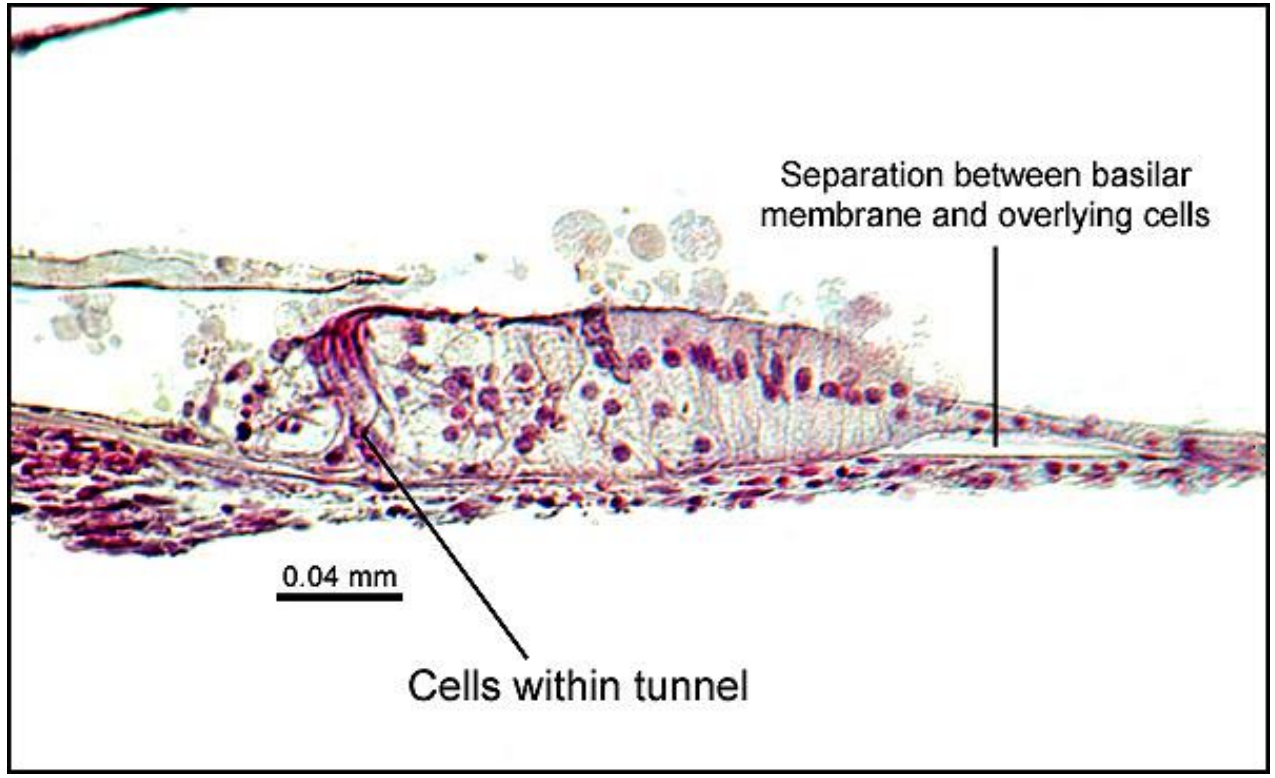
TB Case Number: 727

Gender: Male

Age (yrs.): 20

Otologic Diagnosis:

1. Alport's syndrome
2. Otosclerosis, without stapes fixation, bilateral



Alport Syndrome

Title: Alport syndrome

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Alport Syndrome

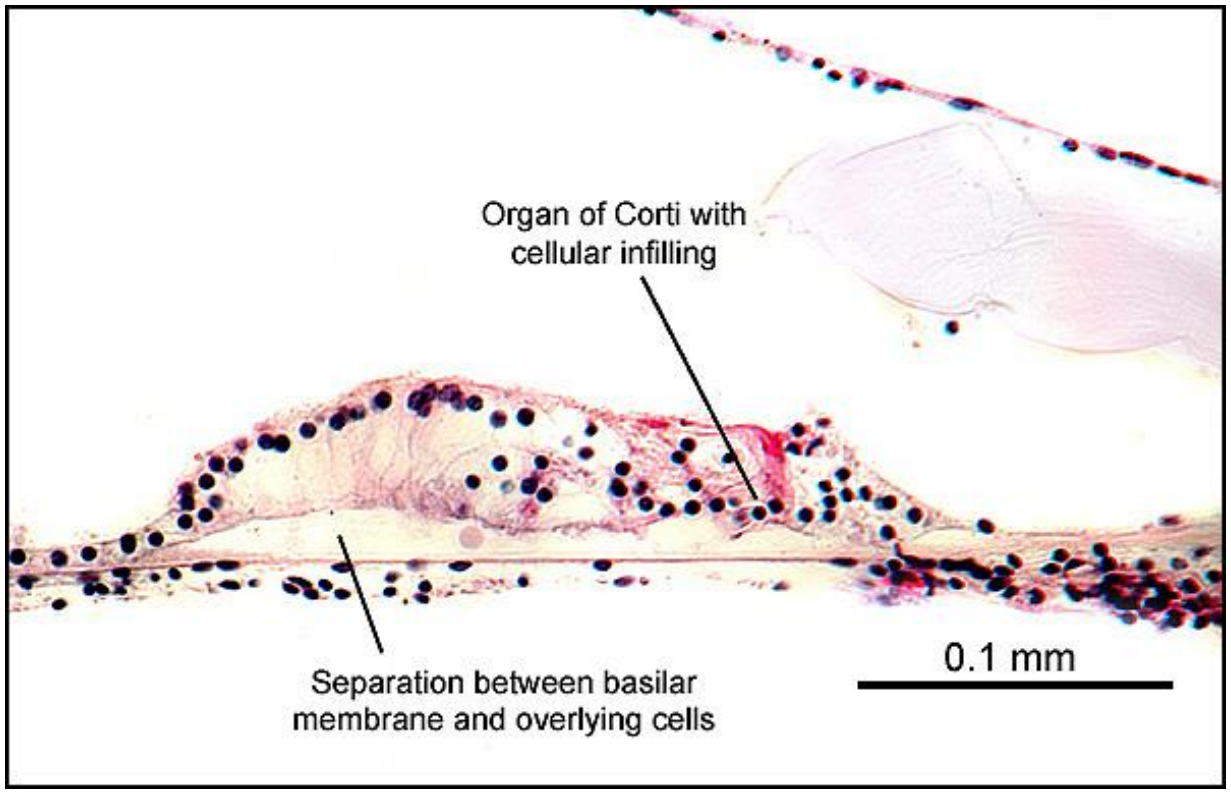
TB Case Number: 801

Gender: Male

Age (yrs.): 50

Otologic Diagnosis:

1. Alport's syndrome, with hearing loss by history (no audiogram) and renal failure
2. Lymphomatoid granulomatosis as manifested by perivascular infiltrates of the vessels of the internal auditory canal and infiltrates in the nerve trunks.



Nieman Pick Syndrome, R-221

Title: Nieman Pick Syndrome, R-221

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mucopolysaccharidosis

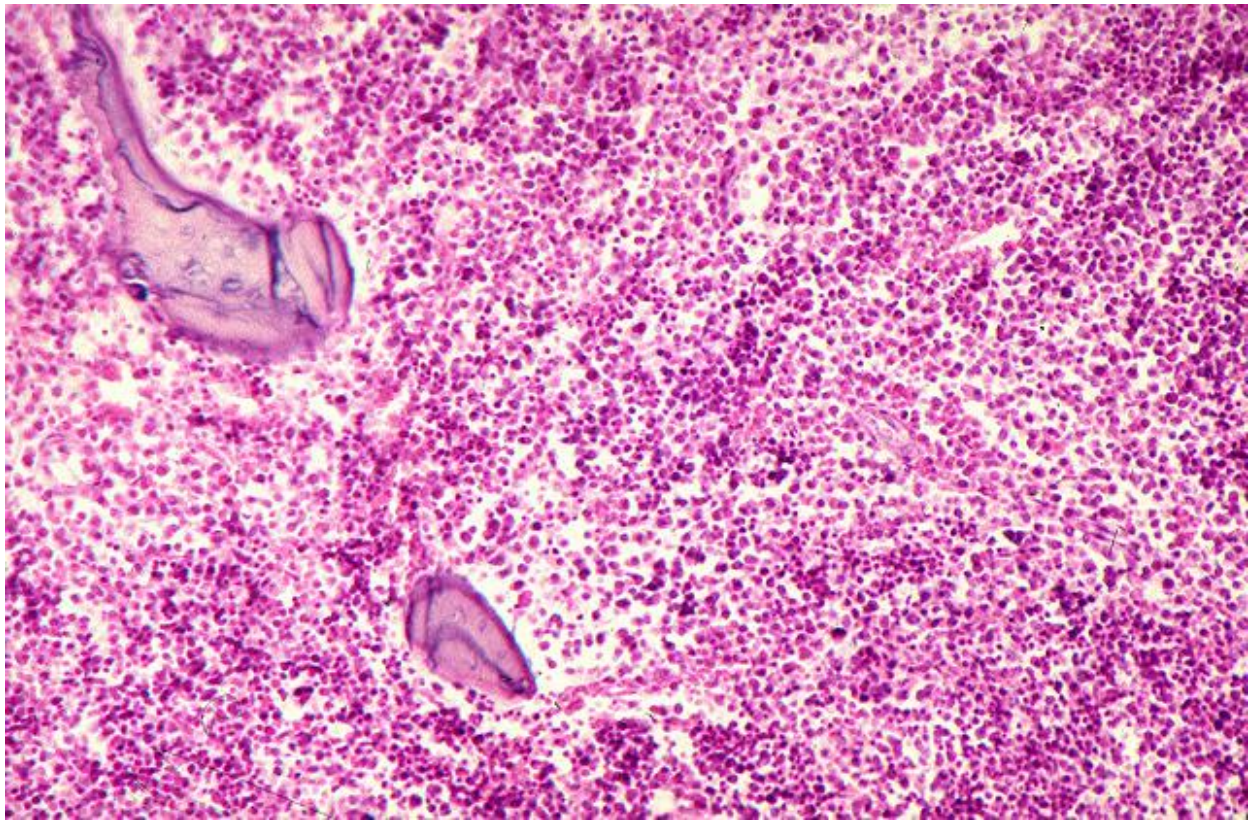
TB Case Number: 557

Gender: Male

Age (yrs.): 1

Otologic Diagnosis:

1. Niemann-Pick disease, Type A
2. Mesenchyme, unresolved, modified, tympanic cavity, right
3. Incudostapedial articulation, hydrarthrosis, right
4. Mastoid, retarded development
5. Perilymph, cellular syncytium, vestibule, right



Nieman Pick Syndrome, R-221

Title: Nieman Pick syndrome, R-221

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mucopolysaccharidosis

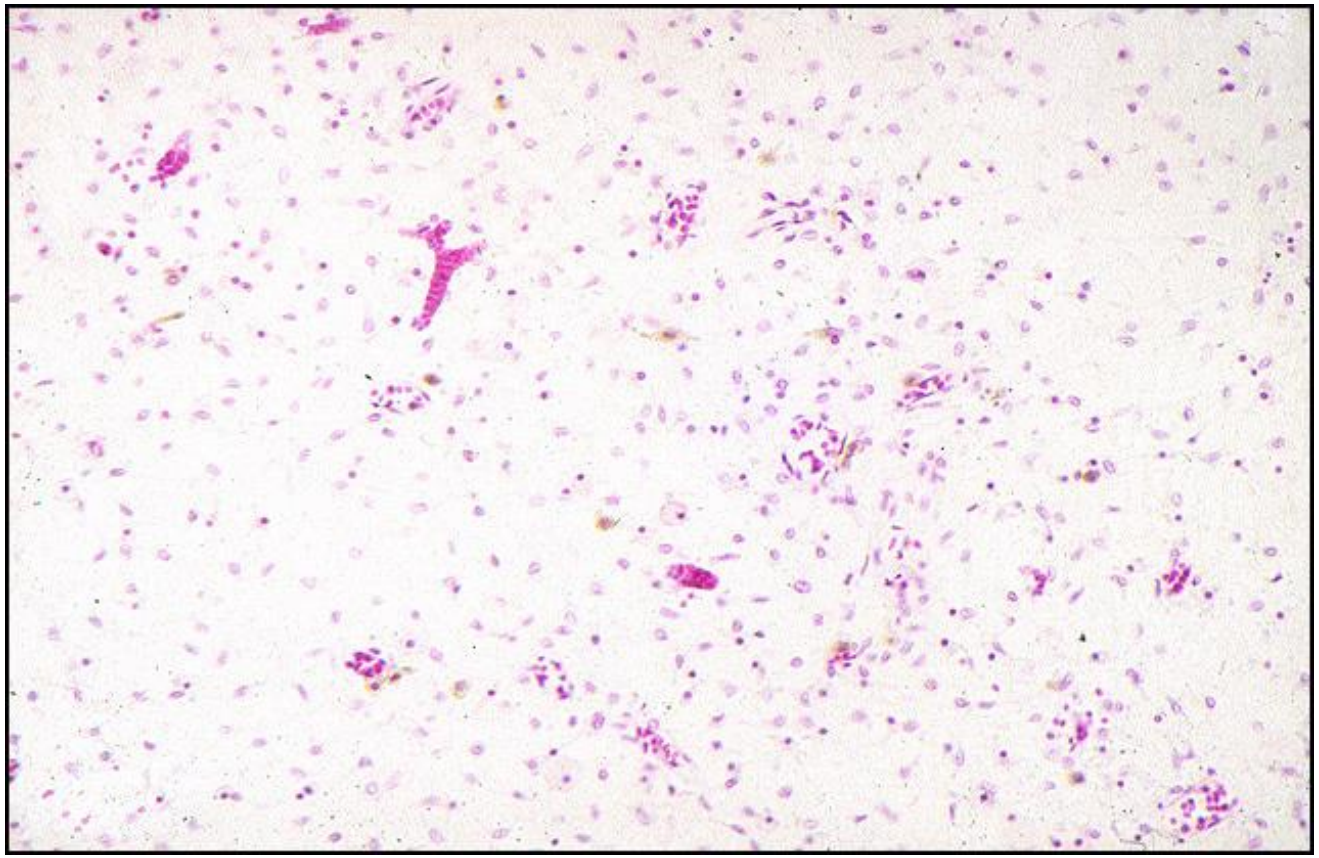
TB Case Number: 557

Gender: Male

Age (yrs.): 1

Otologic Diagnosis:

1. Niemann-Pick disease, Type A
2. Mesenchyme, unresolved, modified, tympanic cavity, right
3. Incudostapedial articulation, hydrarthrosis, right
4. Mastoid, retarded development
5. Perilymph, cellular . syncytium, vestibule, right



Nieman Pick Syndrome, R-221

Title: Nieman Pick Syndrome, R-221

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mucopolysaccharidosis

TB Case Number: 557

Gender: Male

Age (yrs.): 1

Otologic Diagnosis:

1. Niemann-Pick disease, Type A
2. Mesenchyme, unresolved, modified, tympanic cavity, right
3. Incudostapedial articulation, hydrarthrosis, right
4. Mastoid, retarded development
5. Perilymph, cellular syncytium, vestibule, right

Nieman Pick Syndrome, R-81

Title: Nieman Pick Syndrome, R-81

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mucopolysaccharidosis

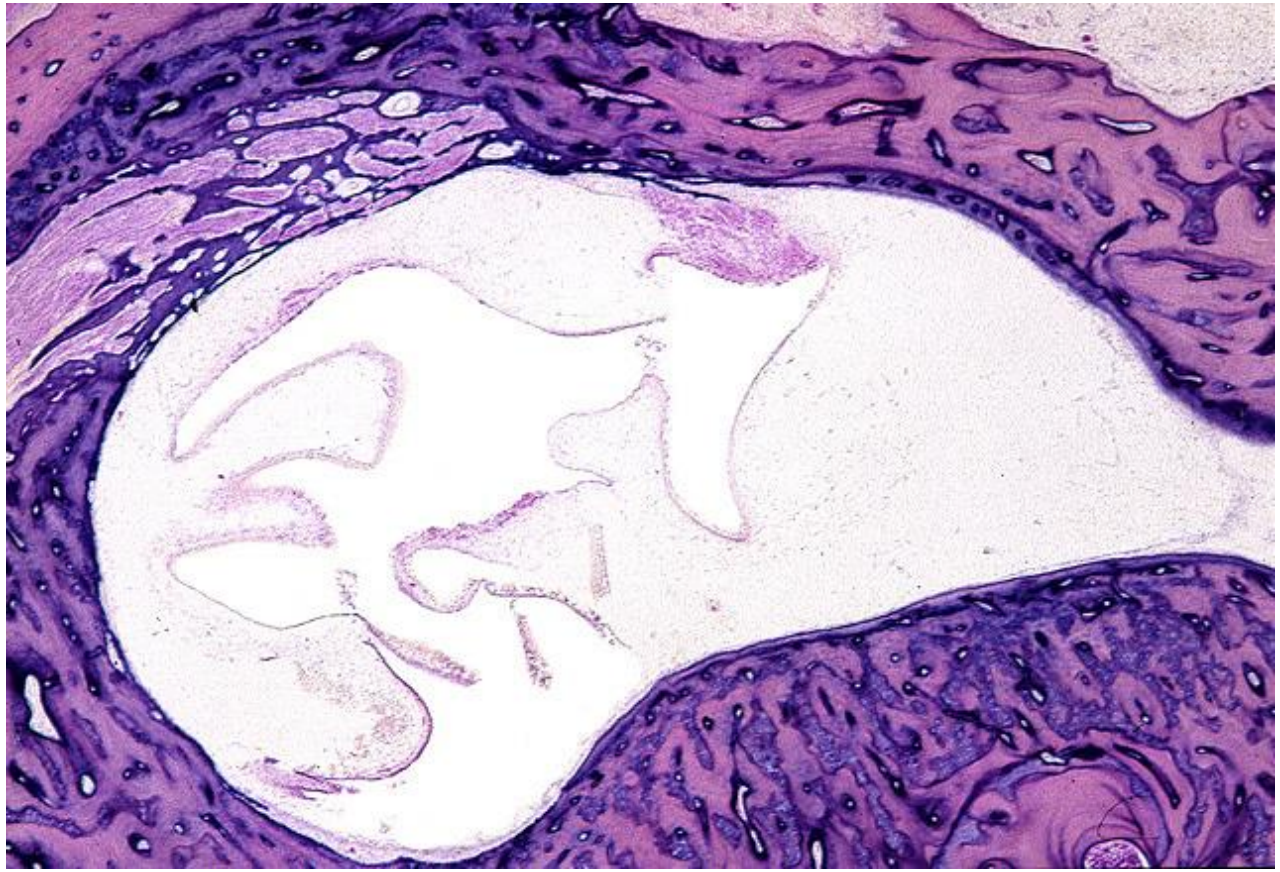
TB Case Number: 557

Gender: Male

Age (yrs.): 1

Otologic Diagnosis:

1. Niemann-Pick disease, type A
2. Mesenchyme, unresolved, modified, tympanic cavity, right
3. Incudostapedial articulation, hydrarthrosis, right
4. Mastoid, retarded development
5. Perilymph, cellular syncytium, vestibule, right



Mucolipidosis, R-351

Title: Mucolipidosis, R-351

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mucopolysaccharidosis

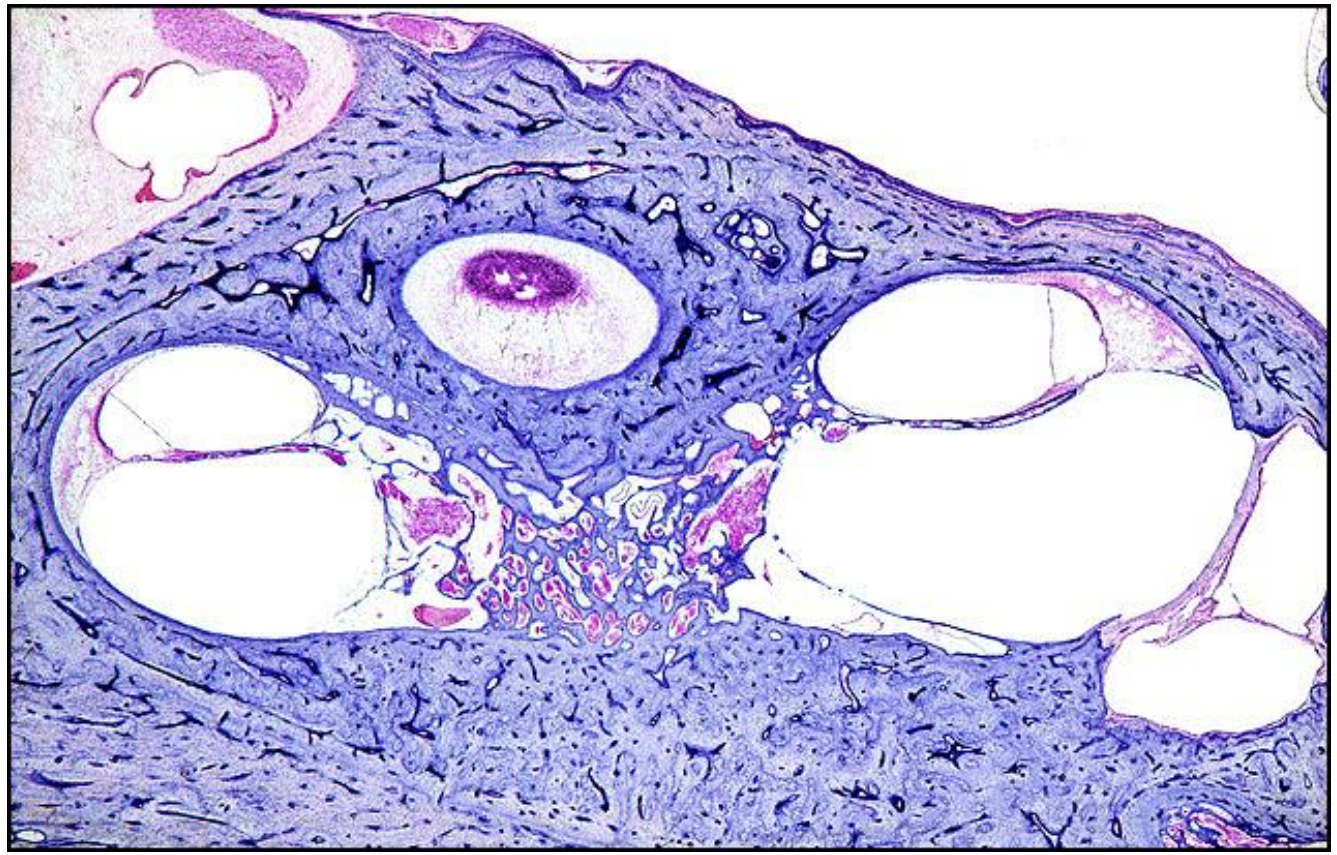
TB Case Number: 803

Gender: Male

Age (yrs.): 24

Otologic Diagnosis:

1. Mucolipidosis type IV (clinical and autopsy diagnosis)
2. Cystic formations, spiral ligament, basal turn, significance unknown, right
3. Middle and inner ears, normal except for above



Mucolipidosis, R-351

Title: Mucolipidosis, R-351

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mucopolysaccharidosis

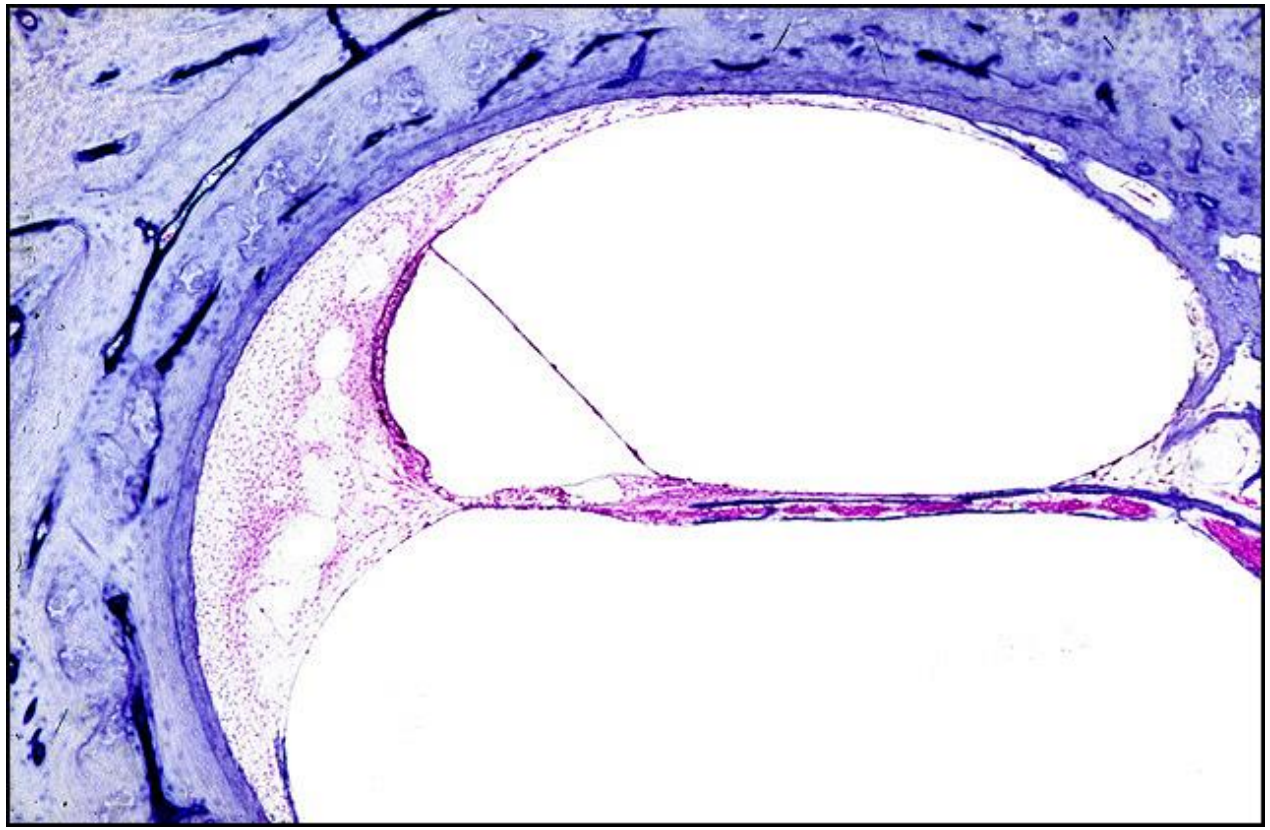
TB Case Number: 803

Gender: Male

Age (yrs.): 24

Otologic Diagnosis:

1. Mucolipidosis type IV (clinical and autopsy diagnosis)
2. Cystic formations, spiral ligament, basal turn, significance unknown, right
3. Middle and inner ears, normal except for above



Mucolipidosis, R-351

Title: Mucolipidosis, R-351

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Mucopolysaccharidosis

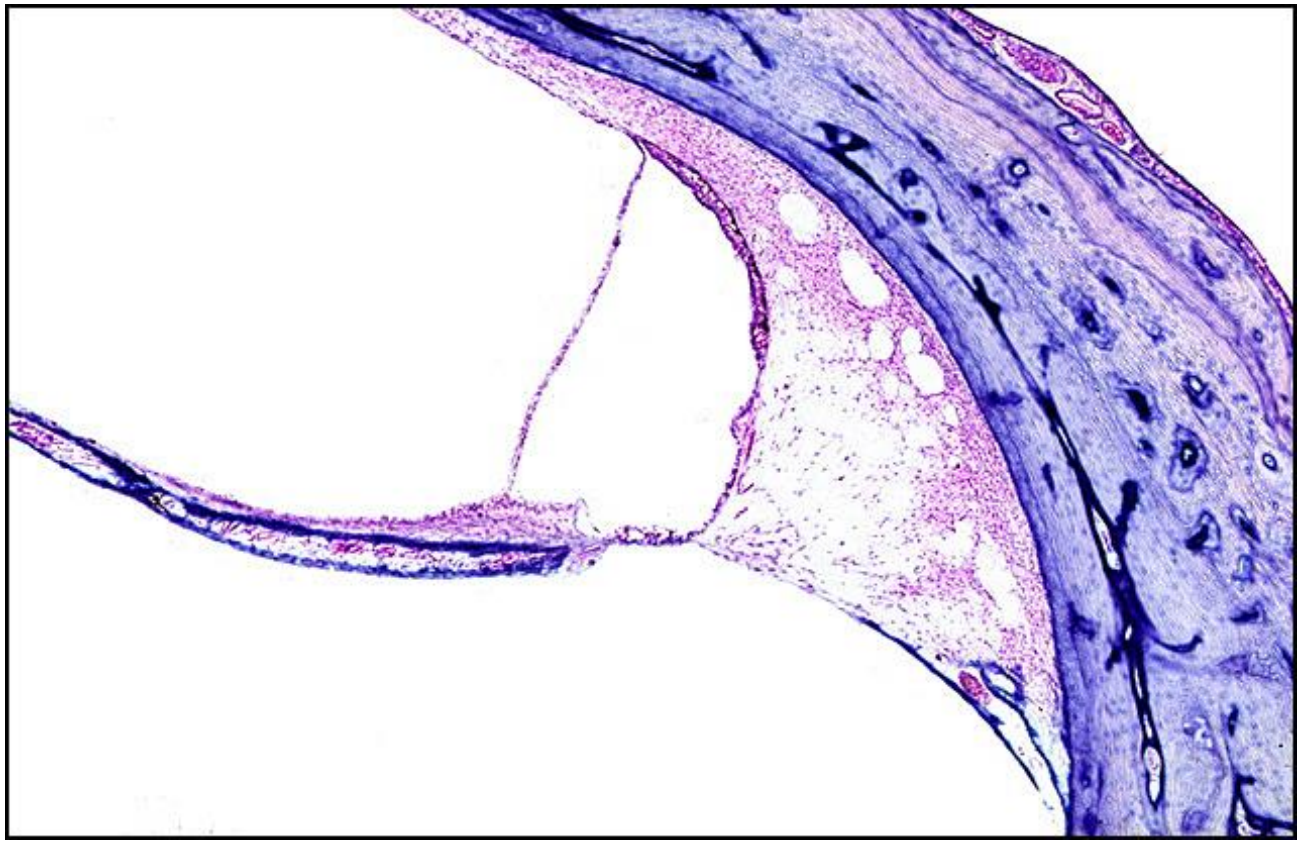
TB Case Number: 803

Gender: Male

Age (yrs.): 24

Otologic Diagnosis:

1. Mucolipidosis type IV (clinical and autopsy diagnosis)
2. Cystic formations, spiral ligament, basal turn, significance unknown, right
3. Middle and inner ears, normal except for above



Norrie Disease, R-76

Title: Norrie Disease, R-76

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Norrie's Disease

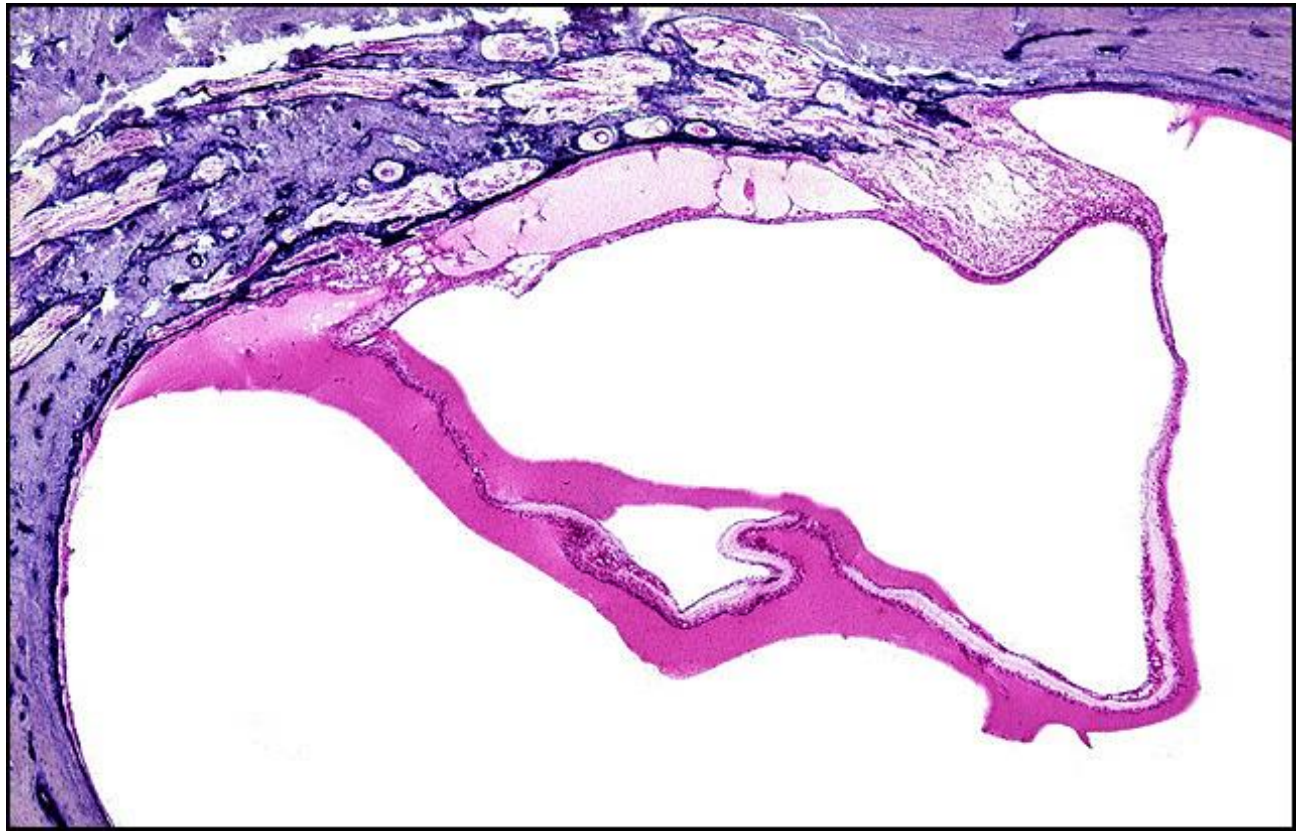
TB Case Number: 532

Gender: Male

Age (yrs.): 77

Otologic Diagnosis:

1. Norrie’s syndrome, characterized by profound hearing loss, bilateral
2. Severe degeneration of the organ of Corti, tectorial membrane, stria vascularis, and cochlear neurons, right ear Arachnoid cyst of the internal auditory canal.



Norrie Disease, R-91

Title: Norrie Disease, R-91

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Norrie's Disease

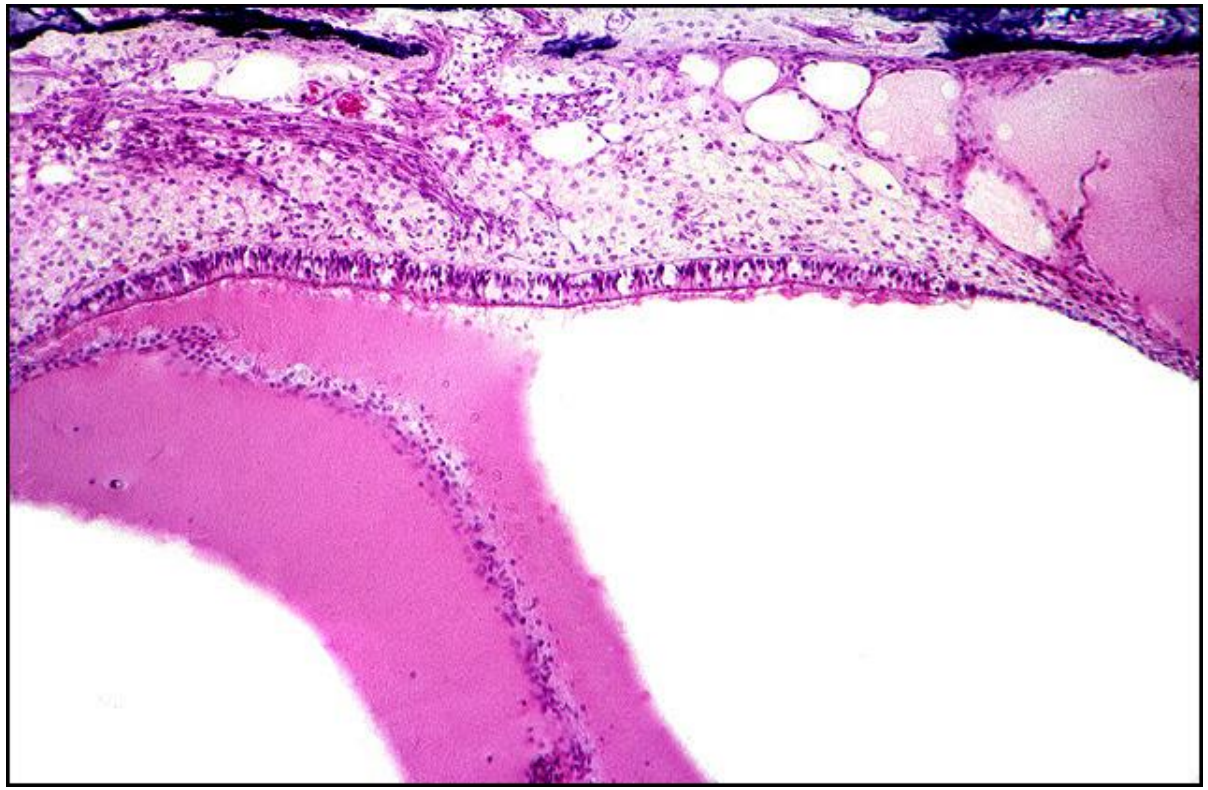
TB Case Number: 532

Gender: Male

Age (yrs.): 77

Otologic Diagnosis:

1. Norrie’s syndrome, characterized by profound hearing loss, bilateral
2. Severe degeneration of the organ of Corti, tectorial membrane, stria vascularis, and cochlear neurons, right ear Arachnoid cyst of the internal auditory canal.



Norrie Disease, R-181

Title: Norrie Disease, R-181

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Norrie's Disease

TB Case Number: 532

Gender: Male

Age (yrs.): 77

Otologic Diagnosis:

1. Norrie’s syndrome, characterized by profound hearing loss, bilateral
2. Severe degeneration of the organ of Corti, tectorial membrane, stria vascularis, and cochlear neurons, right ear Arachnoid cyst of the internal auditory canal.

Norrie Disease, R-76

Title: Norrie Disease, R-76

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Norrie's Disease

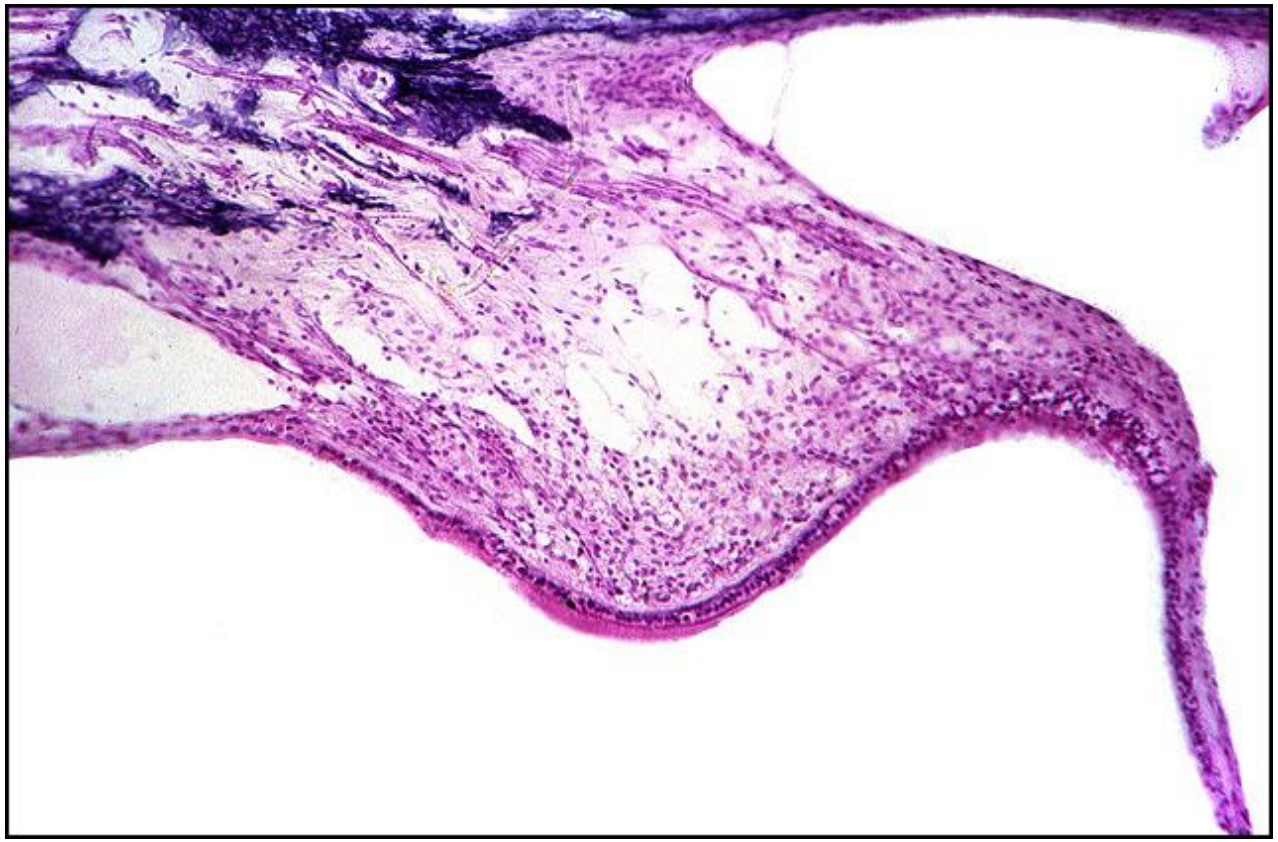
TB Case Number: 532

Gender: Male

Age (yrs.): 77

Otologic Diagnosis:

1. Norrie’s syndrome, characterized by profound hearing loss, bilateral
2. Severe degeneration of the organ of Corti, tectorial membrane, stria vascularis, and cochlear neurons, right ear Arachnoid cyst of the internal auditory canal.



Potter Syndrome, R-276

Title: Potter Syndrome, R-276

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Potter's Syndrome

TB Case Number: 475

Gender: Male

Otologic Diagnosis:

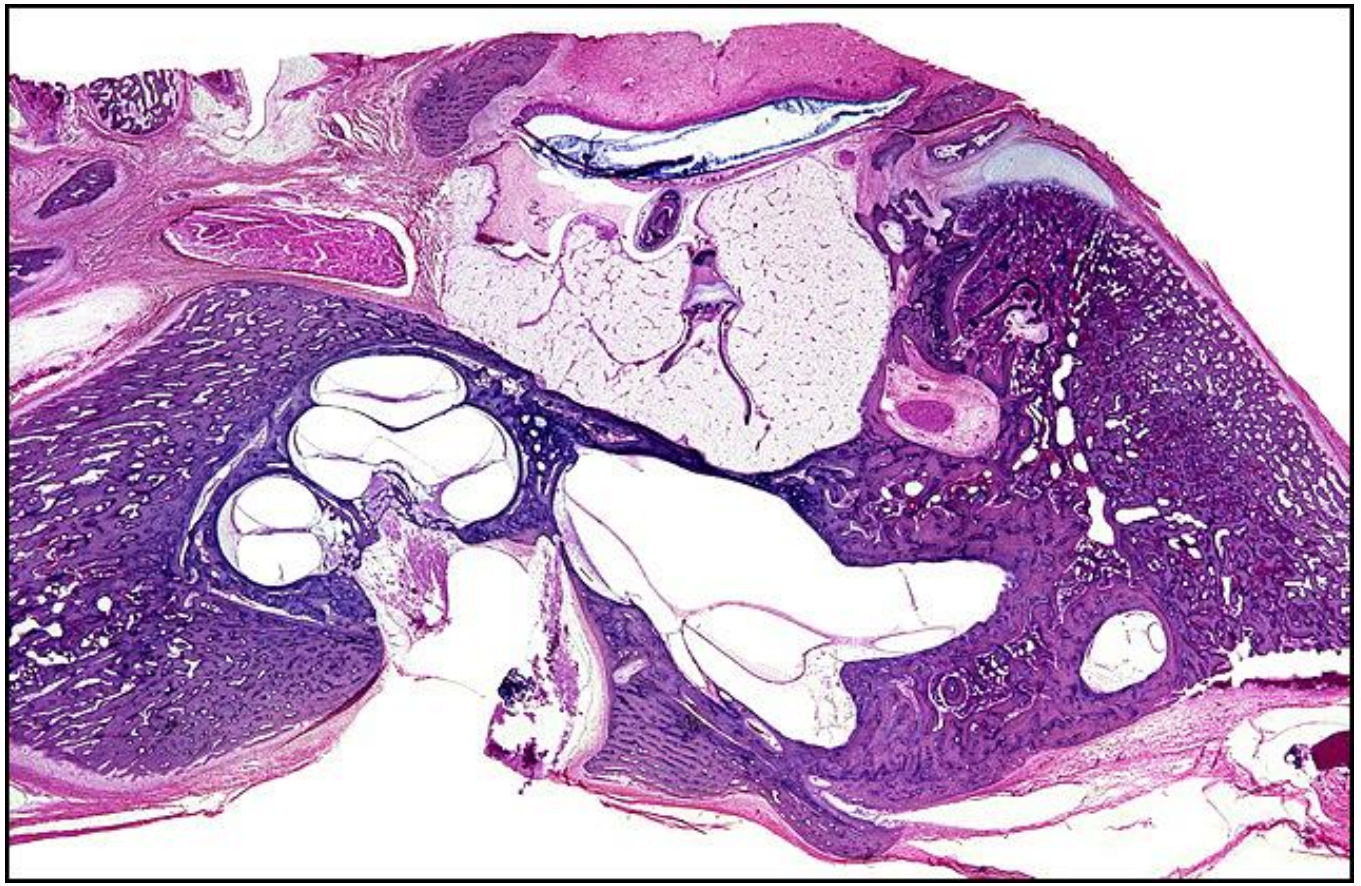
Potter’s syndrome manifested by:

a) Tympanic cavity, delayed cavitation of mesenchyme, bilateral

b) Stapedius muscle, hypoplasia, bilateral

c) Stapedius tendon, absence, bilateral

d) Labyrinth, bony and membranous, normal, bilateral



Potter Syndrome, L-216

Title: Potter Syndrome, L-216

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Potter's Syndrome

TB Case Number: 475

Gender: Male

Otologic Diagnosis:

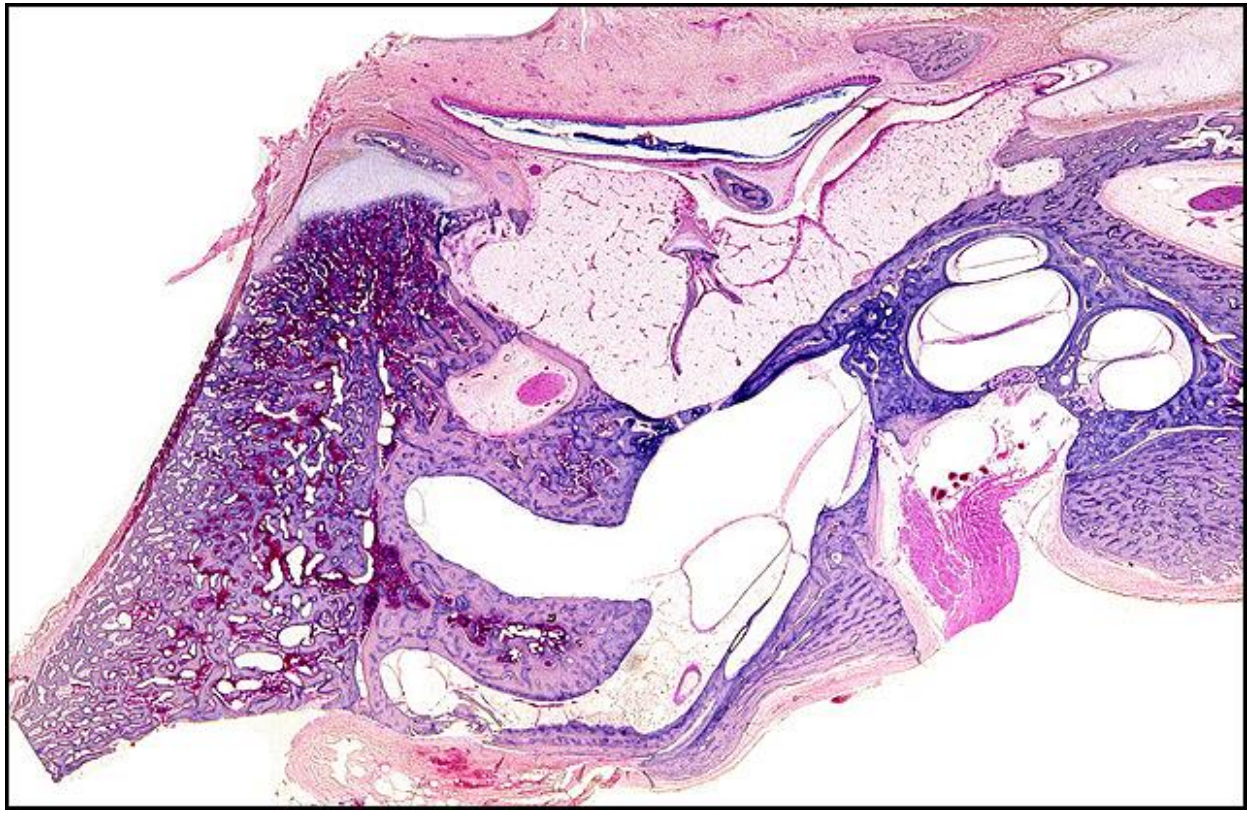
Potter’s syndrome manifested by:

a) Tympanic cavity, delayed cavitation of mesenchyme, bilateral

b) Stapedius muscle, hypoplasia, bilateral

c) Stapedius tendon, absence, bilateral

d) Labyrinth, bony and membranous, normal, bilateral



Multiple Congenital Anomalies, L-141

Title: Multiple Congenital Anomalies, L-141

Chapter: Developmental Defects

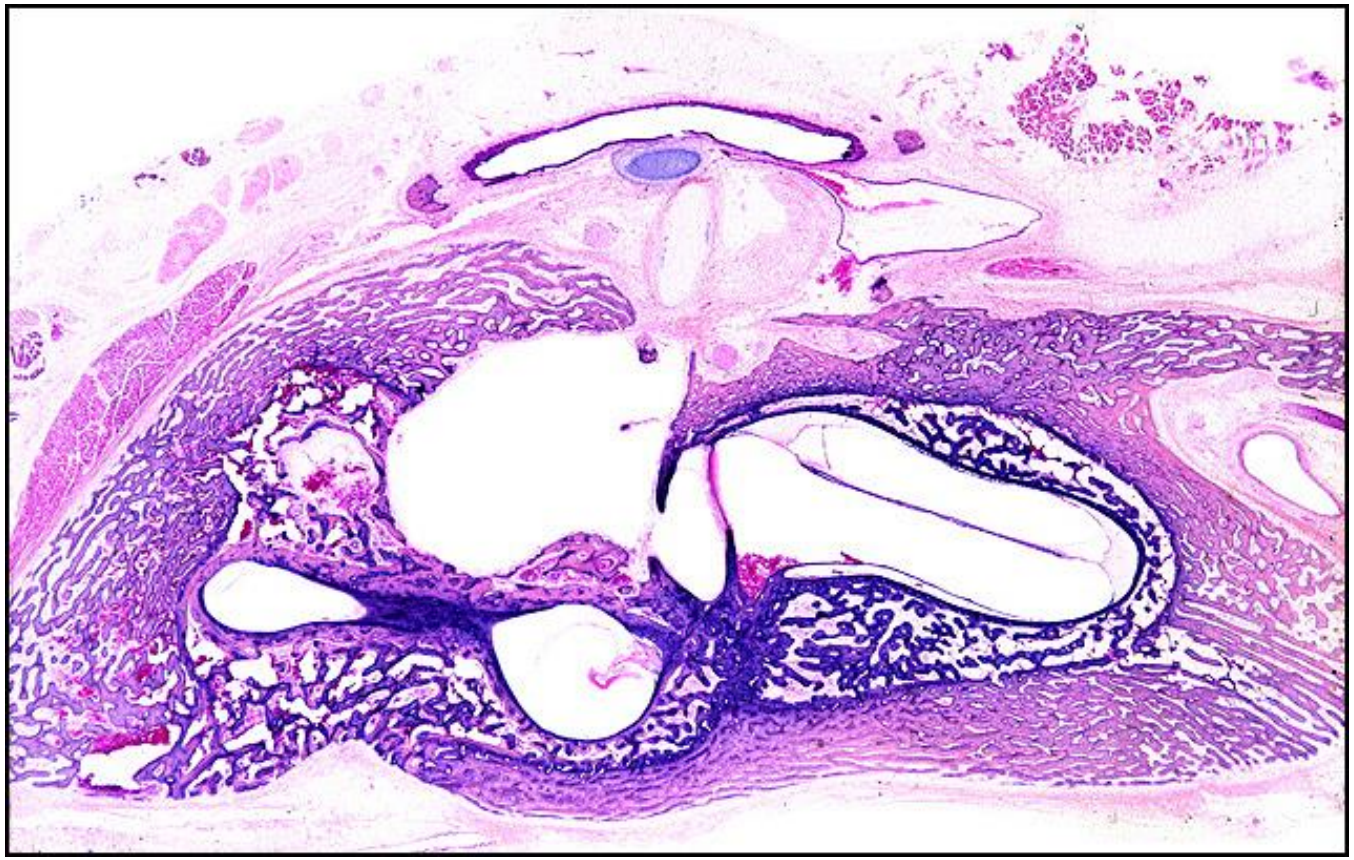
Chapter Section: Syndromic Hearing Loss / Multiple Congenital Anomalies

TB Case Number: 325

Gender: Male

Otologic Diagnosis:

1. Multiple middle ear anomalies including:
    a) Obliteration of the middle ear with mesenchyme and bone
    b) Ossicular anomalies
    c) Congenital cholesteatoma medial to the tympanic membrane
    d) Partial development of the footplate
2. Normal bony and membranous labyrinth



Multiple Congenital Anomalies, L-141

Title: L141 Multiple Congenital Anomalies

Chapter: Developmental Defects

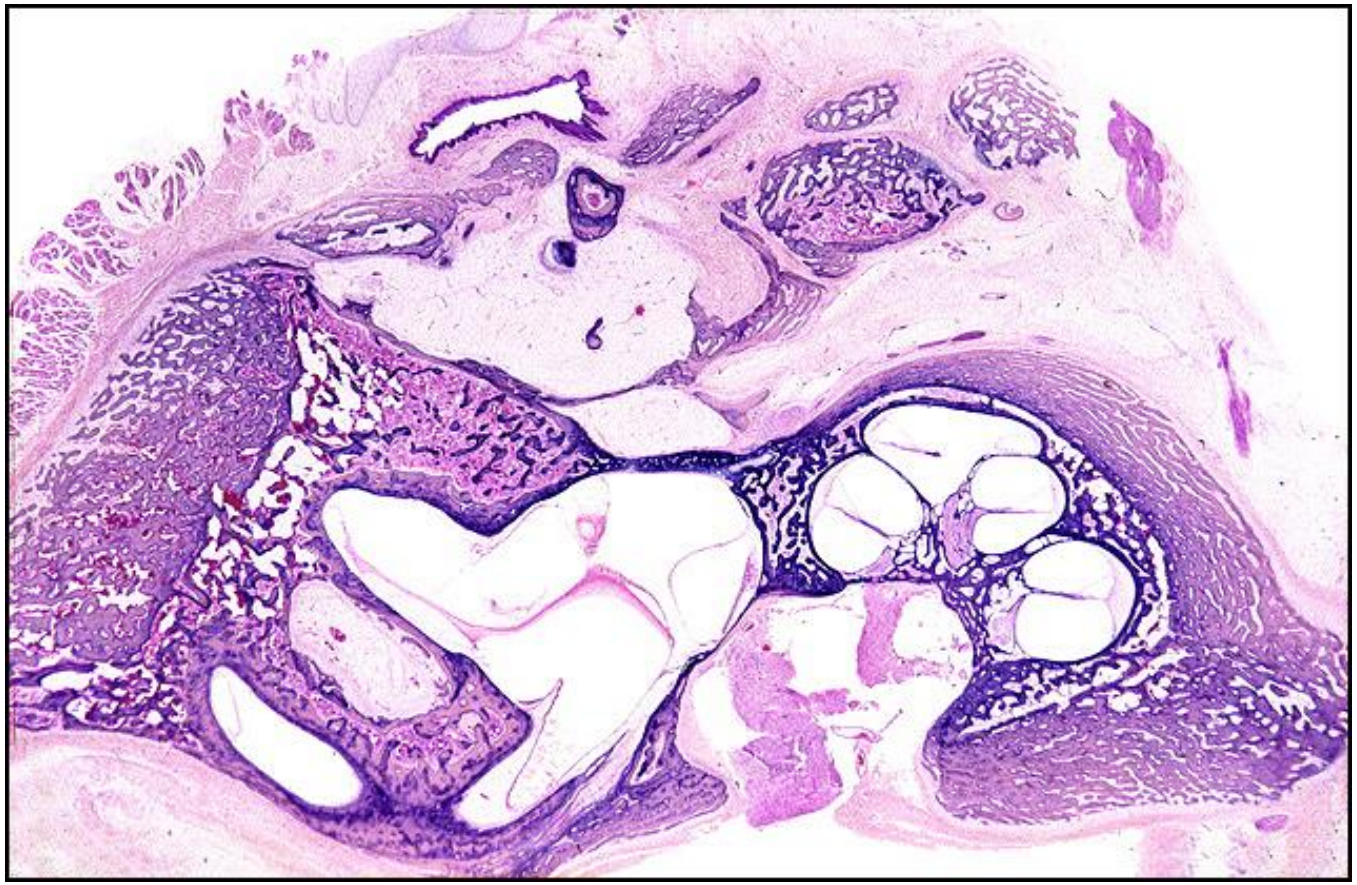
Chapter Section: Syndromic Hearing Loss / Multiple Congenital Anomalies

TB Case Number: 325

Gender: Male

Otologic Diagnosis:

1. Multiple middle ear anomalies including:
    a) Obliteration of the middle ear with mesenchyme and bone
    b) Ossicular anomalies
    c) Congenital cholesteatoma medial to the tympanic membrane
    d) Partial development of the footplate
2. Normal bony and membranous labyrinth



Multiple Congenital Anomalies, L-141

Title: Multiple Congenital Anomalies, L-141

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Multiple Congenital Anomalies

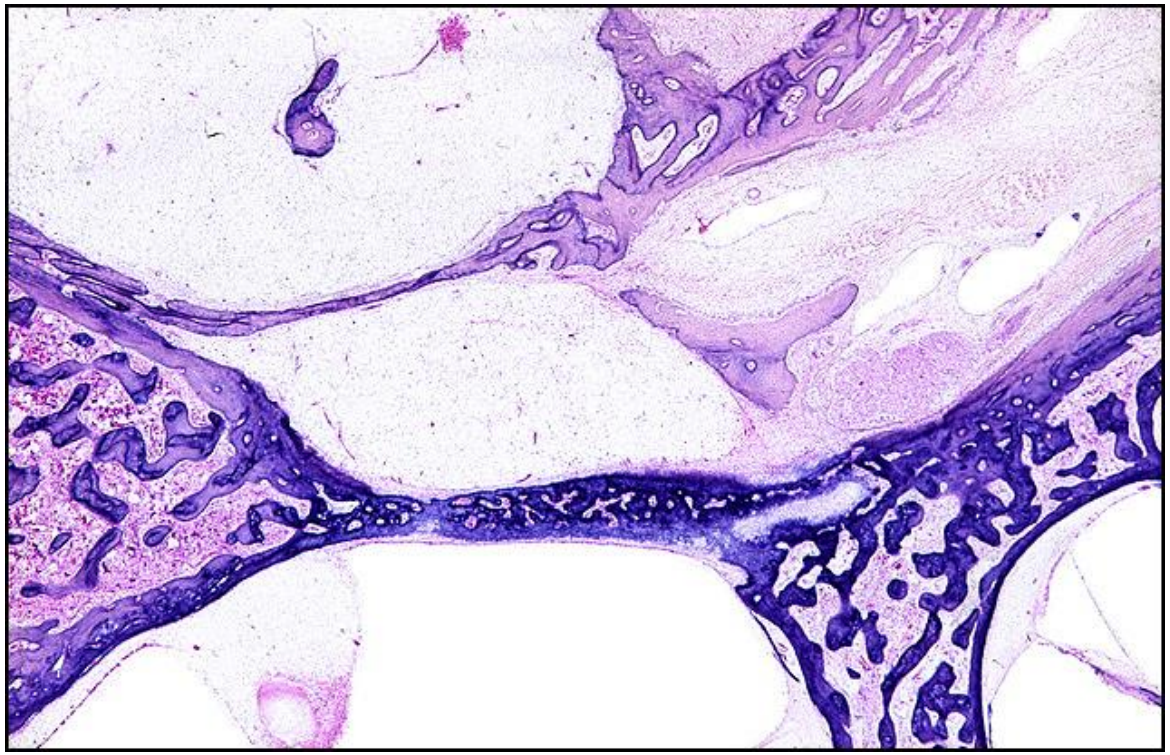
TB Case Number: 325

Comments: Hemorrhage, post surgery, repair of omphalocele, multiple congenital anomalies, prematurity (30 weeks)

Gender: Male

Otologic Diagnosis:

1. Multiple middle ear anomalies including:
    a) Obliteration of the middle ear with mesenchyme and bone
    b) Ossicular anomalies
    c) Congenital cholesteatoma medial to the tympanic membrane
    d) Partial development of the footplate
2. Normal bony and membranous labyrinth



Multiple Anomalies, L-91

Title: Multiple Anomalies, L-91

Chapter: Developmental Defects

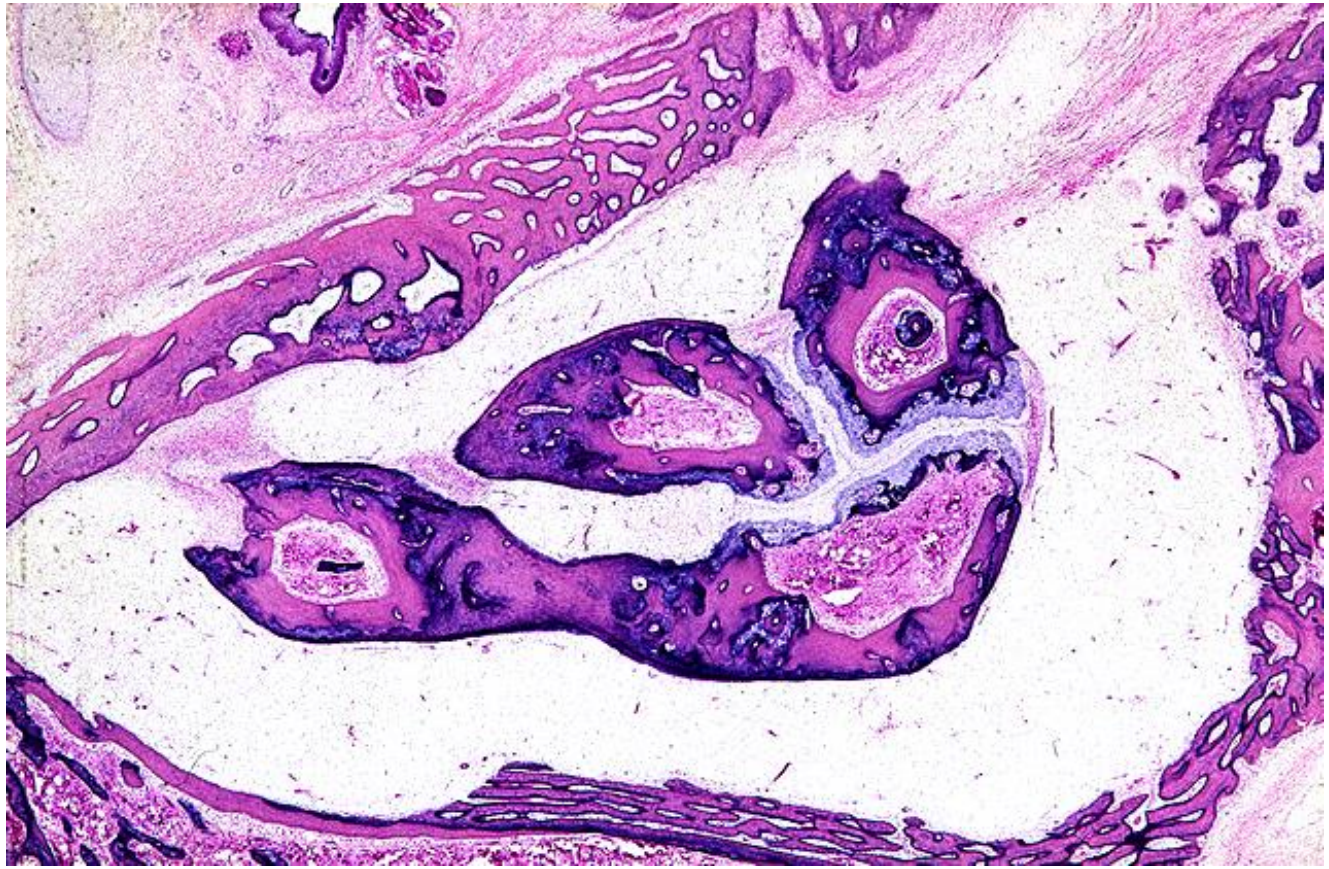
Chapter Section: Syndromic Hearing Loss / Multiple Congenital Anomalies

TB Case Number: 325

Gender: Male

Otologic Diagnosis:

1. Multiple middle ear anomalies including:
    a) Obliteration of the middle ear with mesenchyme and bone
    b) Ossicular anomalies
    c) Congenital cholesteatoma medial to the tympanic membrane
    d) Partial development of the footplate
2. Normal bony and membranous labyrinth



Trisomy 13, L-191

Title: Trisomy 13, L-191

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Trisomy 13 Patau's Syndrome

TB Case Number: 561

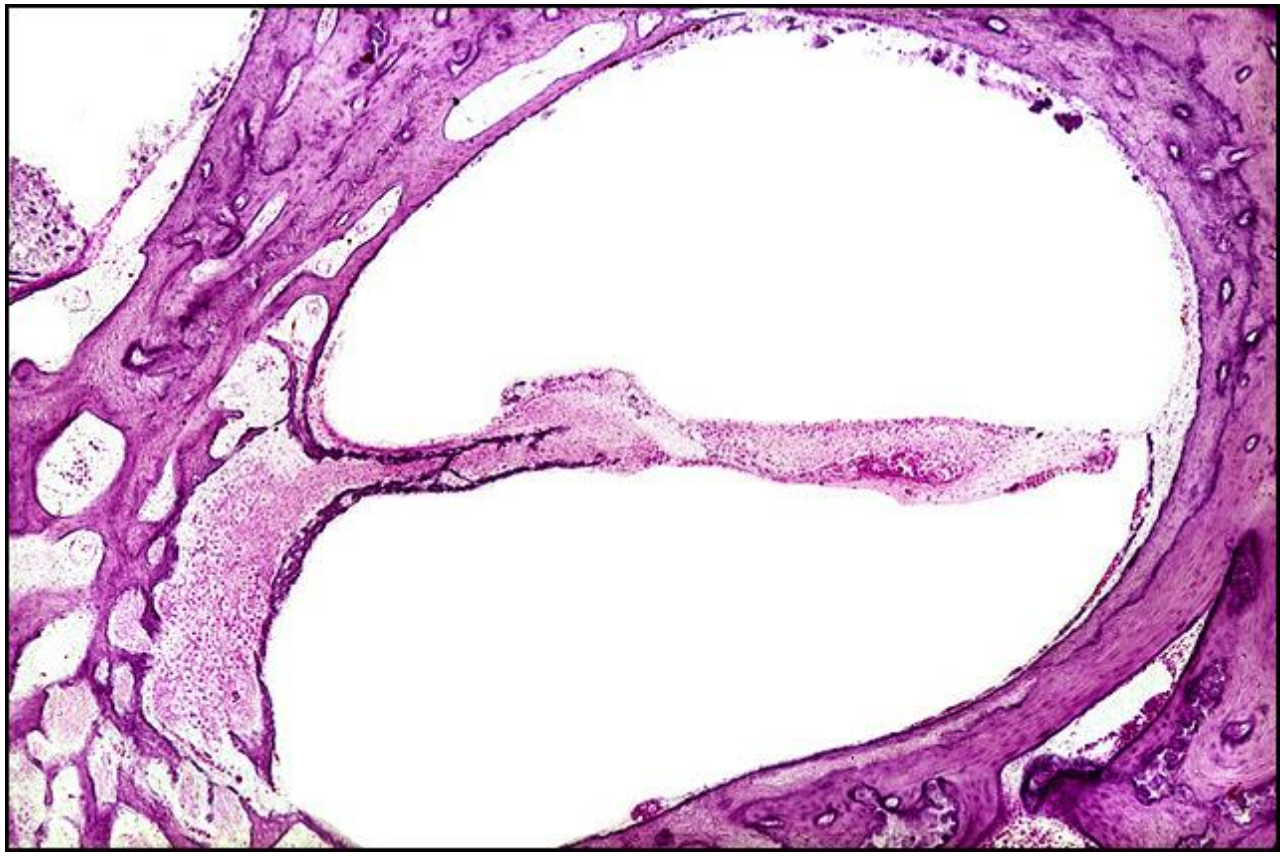
Gender: Female

Otologic Diagnosis:

Trisomy 13-15 syndrome with bilateral otologic malformations including:

a) Hypoplasia of the structures of the cochlear duct, including organ of Corti, stria vascularis, spiral ligament and tectorial membrane

b) Hypoplasia of the saccule



Trisomy 13, R-431

Title: Trisomy 13, R-431

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Trisomy 13 Patau's Syndrome

TB Case Number: 561

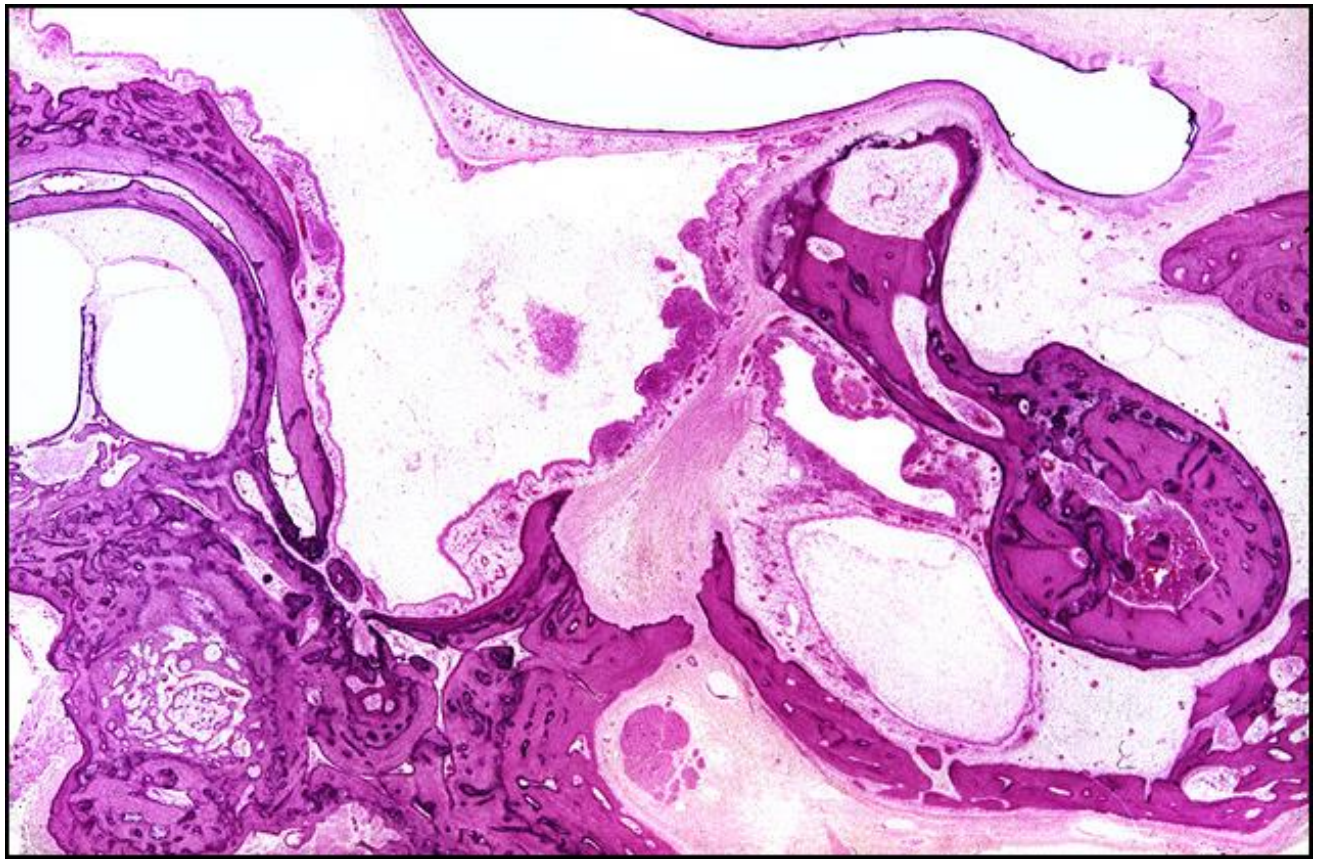
Gender: Female

Otologic Diagnosis:

Trisomy 13-15 syndrome with bilateral otologic malformations including:

a) Hypoplasia of the structures of the cochlear duct, including organ of Corti, stria vascularis, spiral ligament and tectorial membrane

b) Hypoplasia of the saccule



Trisomy 13, R-251

Title: Trisomy 13, R-251

Chapter: Developmental Defects

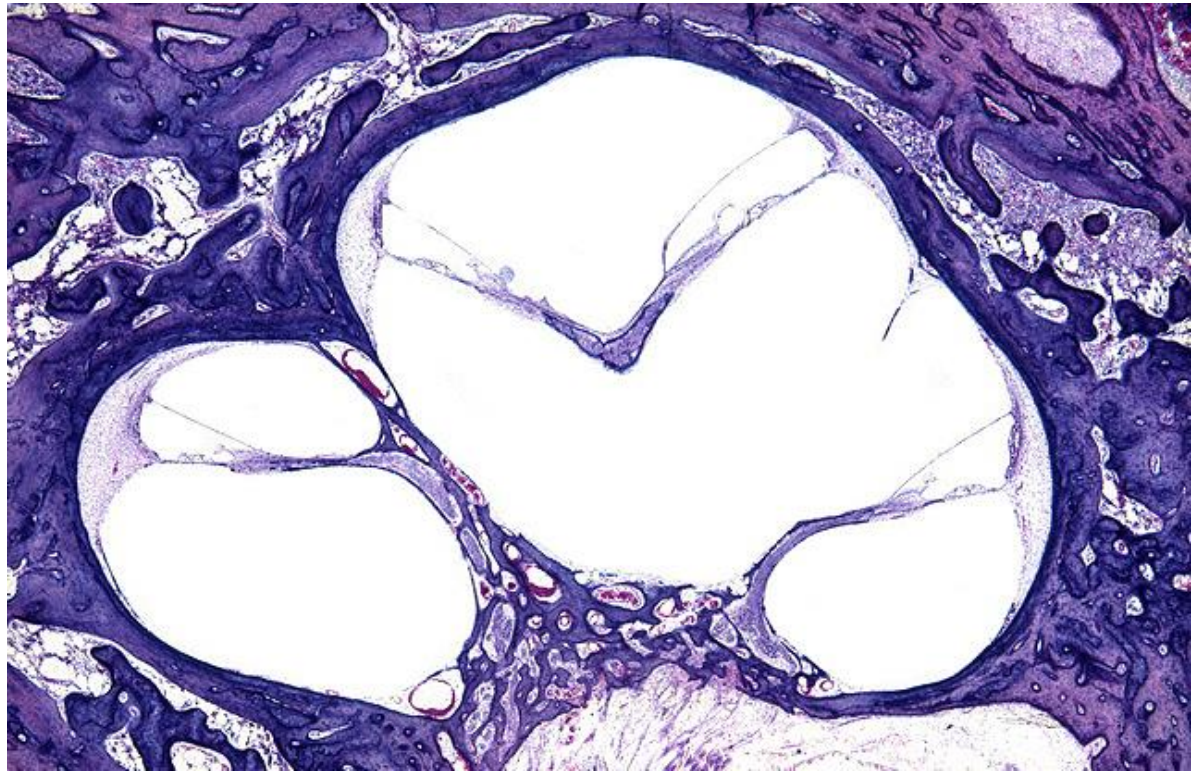
Chapter Section: Syndromic Hearing Loss / Trisomy 13 Patau's Syndrome

TB Case Number: 824

Gender: Male

Otologic Diagnosis:

1. Trisomy 13-15
2. Mondini anomaly, cochlea, bilateral
3. Dysplasia, saccule, bilateral
4. Stria vascularis, cystic areas, bilateral
5. Dysplasia, cochlear duct, bilateral
6. Otitis media, acute, bilateral



Trisomy 13, L-181

Title: Trisomy 13, L-181

Chapter: Developmental Defects

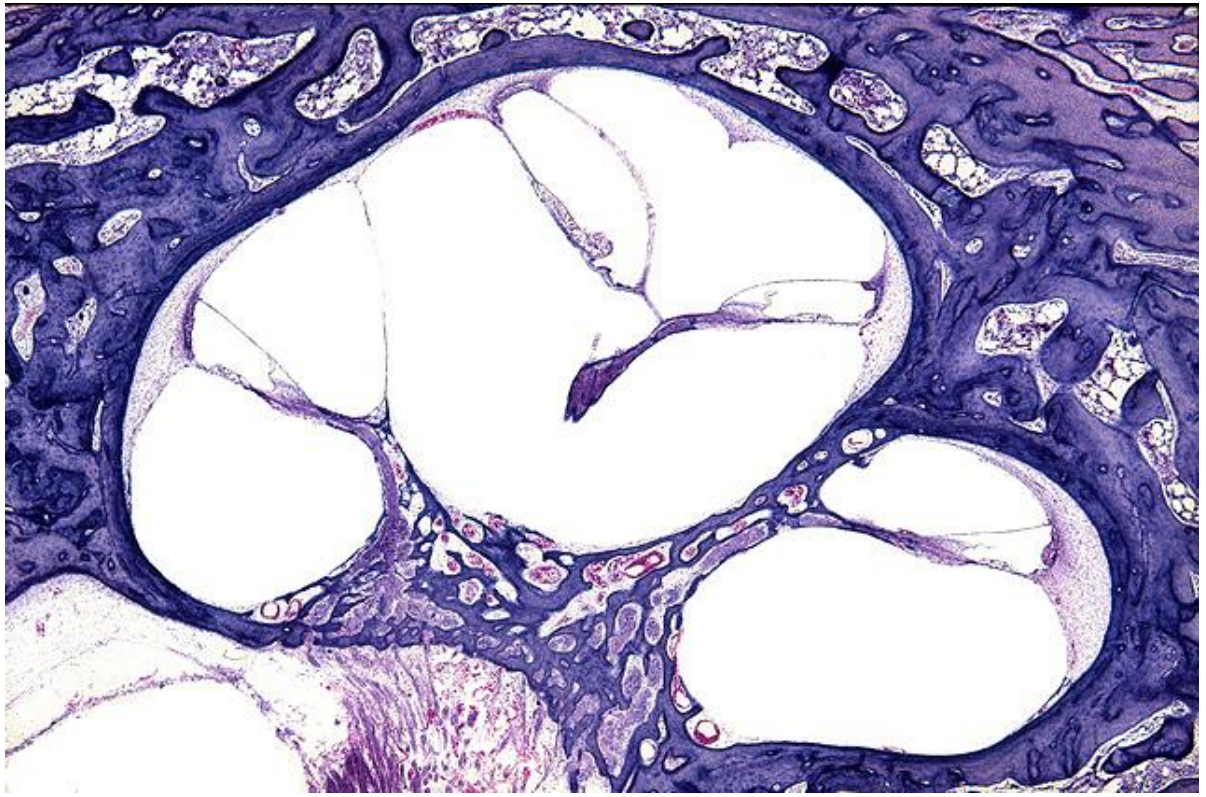
Chapter Section: Syndromic Hearing Loss / Trisomy 13 Patau's Syndrome

TB Case Number: 824

Gender: Male

Otologic Diagnosis:

1. Trisomy 13-15
2. Mondini anomaly, cochlea, bilateral
3. Dysplasia, saccule, bilateral
4. Stria vascularis, cystic areas, bilateral
5. Dysplasia, cochlear duct, bilateral
6. Otitis media, acute, bilateral



Trisomy 13, R-161

Title: Trisomy 13, R-161

Chapter: Developmental Defects

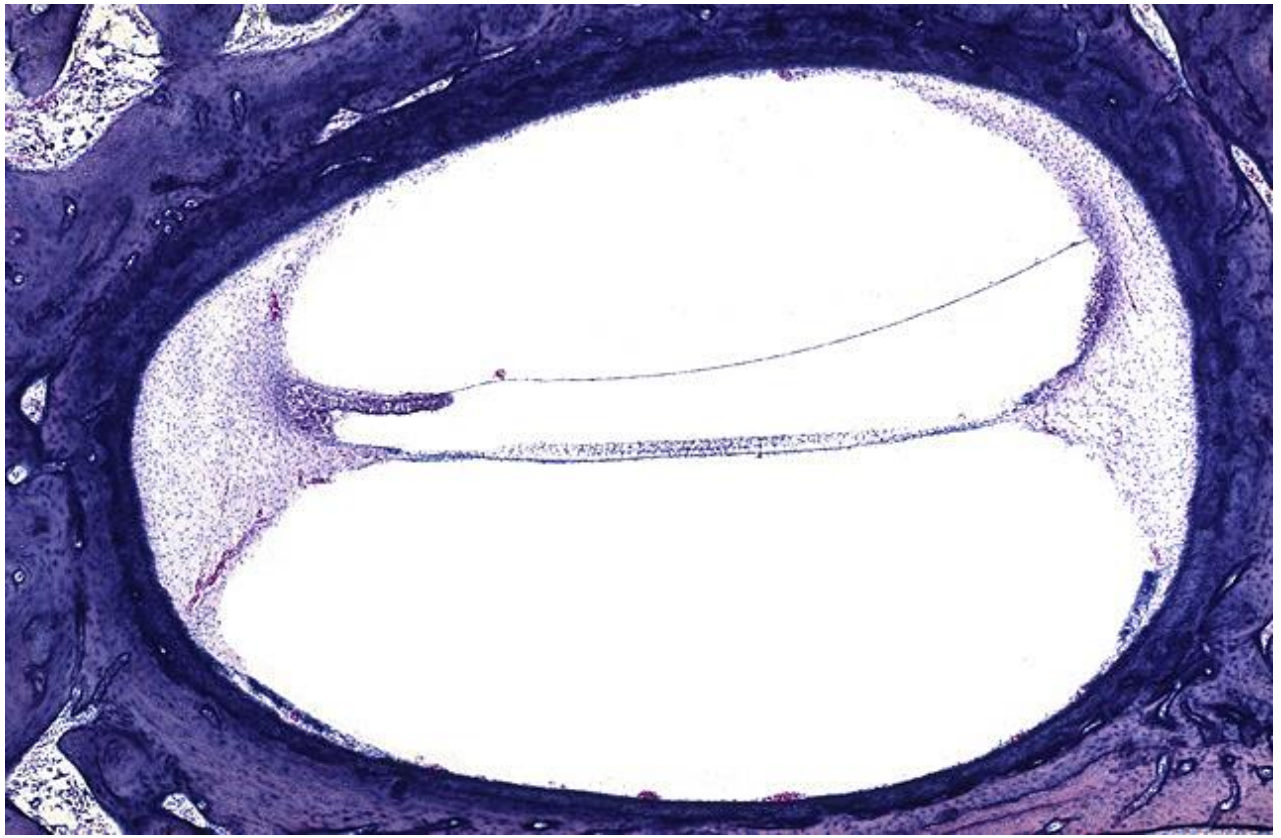
Chapter Section: Syndromic Hearing Loss / Trisomy 13 Patau's Syndrome

TB Case Number: 824

Gender: Male

Otologic Diagnosis:

1. Trisomy 13-15
2. Mondini anomaly, cochlea, bilateral
3. Dysplasia, saccule, bilateral
4. Stria vascularis, cystic areas, bilateral
5. Dysplasia, cochlear duct, bilateral
6. Otitis media, acute, bilateral



Trisomy 18, L-171

Title: Trisomy 18, L-171

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Trisomy 18 Edward's Syndrome

TB Case Number: 595

Gender: Male

Otologic Diagnosis:

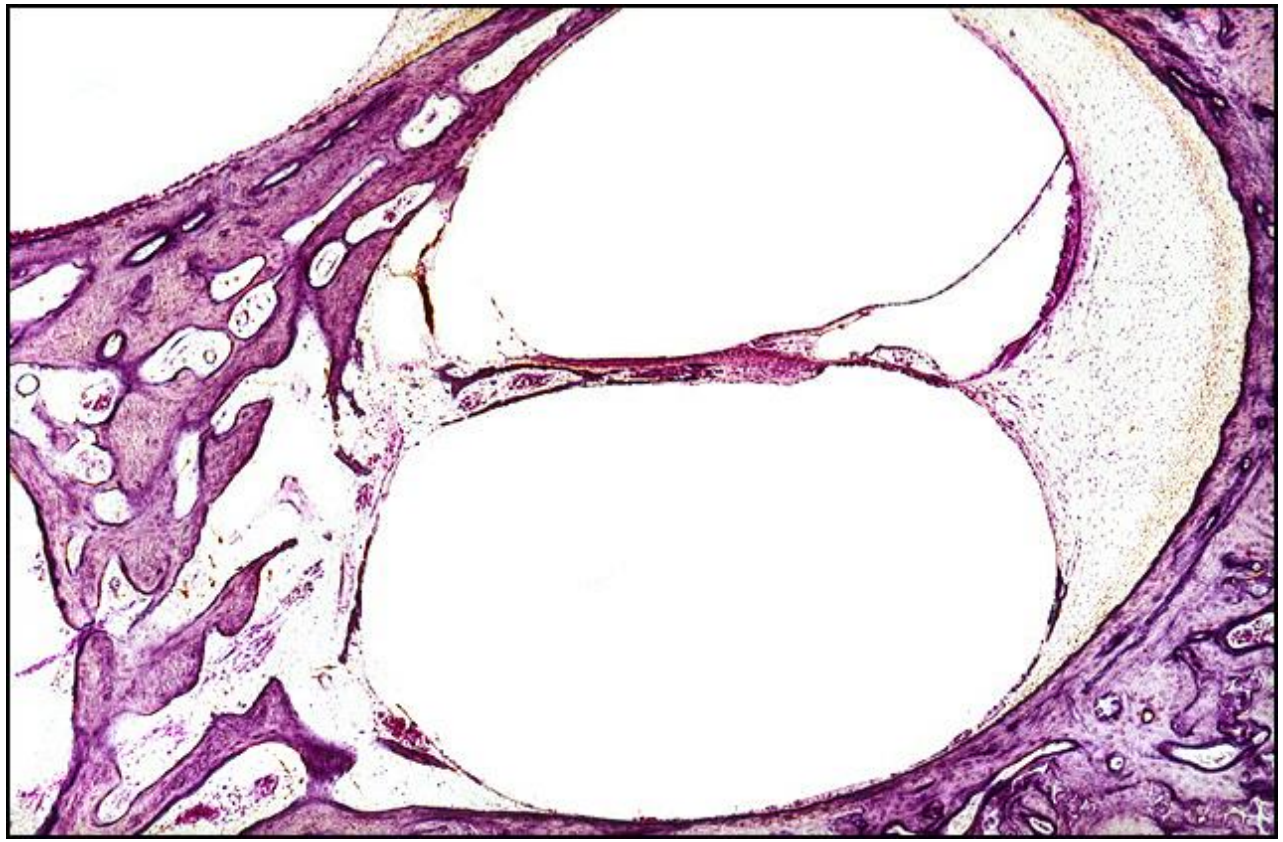
Trisomy-18 syndrome:

a) Malleus, slightly deformed, right

b) Incus, slightly deformed, right

c) Tensor tympani muscle, unusual anatomical course, divided into 2bundles in separate bony canals, right

d) Bony modiolus



Trisomy 18, L-131

Title: Trisomy 18, L-131

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Trisomy 18 Edward's Syndrome

TB Case Number: 595

Gender: Male

Otologic Diagnosis:

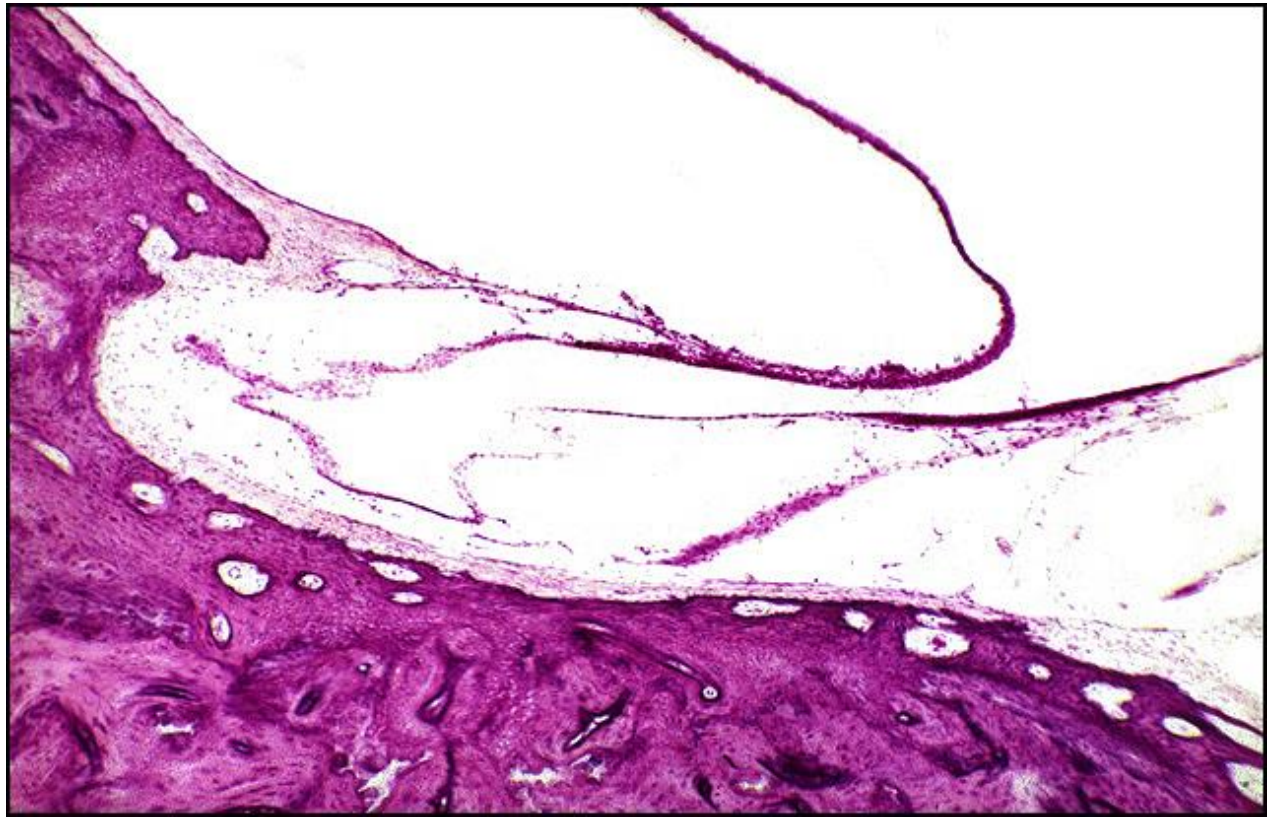
Trisomy-18 syndrome:

a) Malleus, slightly deformed, right

b) Incus, slightly deformed, right

c) Tensor tympani muscle, unusual anatomical course, divided into 2bundles in separate bony canals, right

d) Bony modiolus



Trisomy 18, L-161

Title: Trisomy 18, L-161

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Trisomy 18 Edward's Syndrome

TB Case Number: 595

Gender: Male

Otologic Diagnosis:

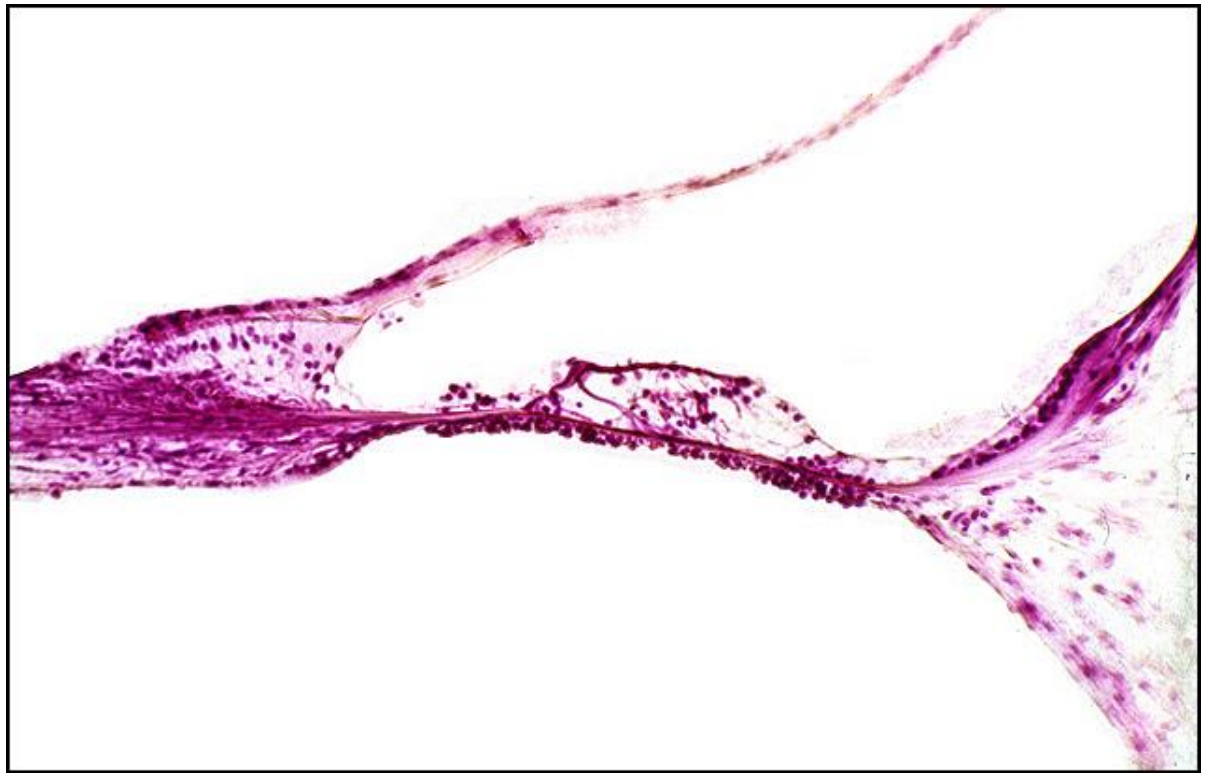
Trisomy-18 syndrome:

a) Malleus, slightly deformed, right

b) Incus, slightly deformed, right

c) Tensor tympani muscle, unusual anatomical course, divided into 2bundles in separate bony canals, right

d) Bony modiolus



Trisomy 18, L-171

Title: Trisomy 18, L-171

Chapter: Developmental Defects

Chapter Section: Syndromic Hearing Loss / Trisomy 18 Edward's Syndrome

TB Case Number: 595

Gender: Male

Otologic Diagnosis:

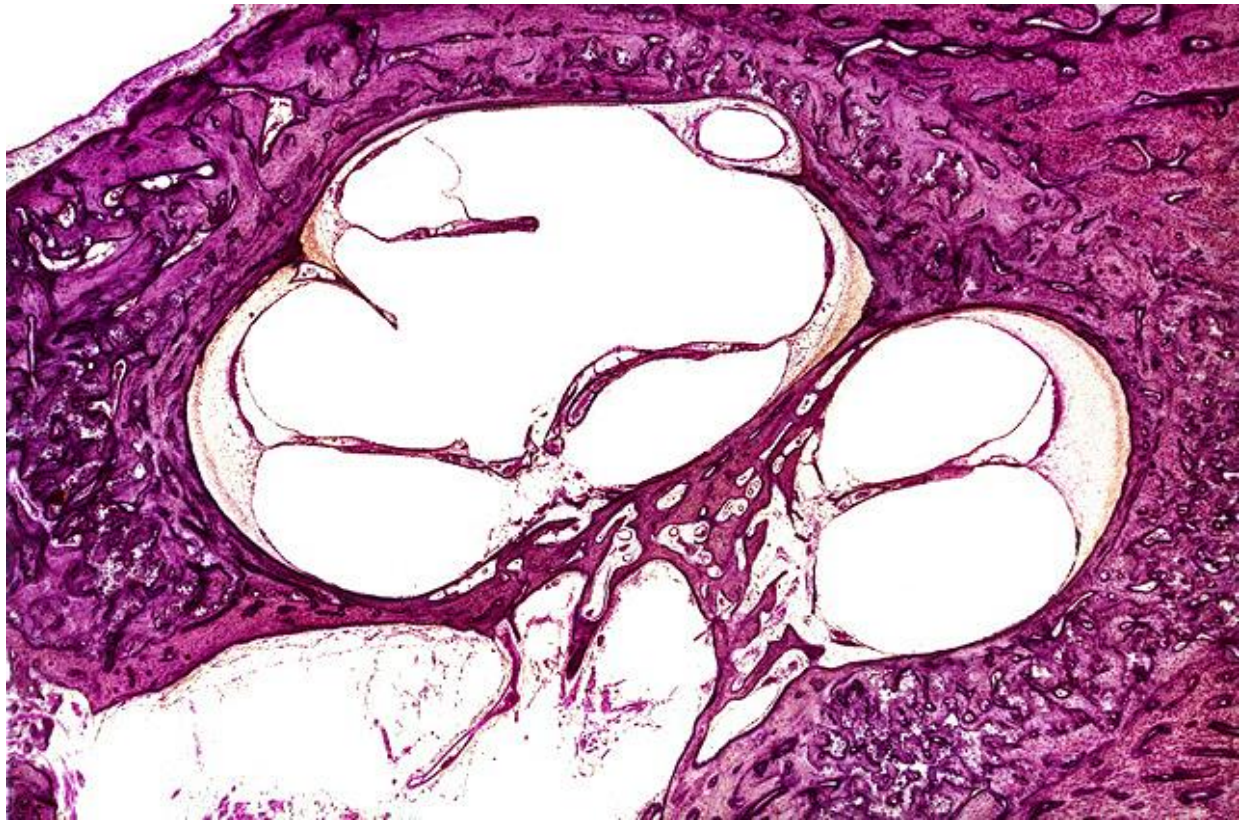
Trisomy-18 syndrome:

a) Malleus, slightly deformed, right

b) Incus, slightly deformed, right

c) Tensor tympani muscle, unusual anatomical course, divided into 2bundles in separate bony canals, right

d) Bony modiolus



Axial Mesodermal Dysplasia, R-351

Title: Axial Mesodermal Dysplasia, R-351

Chapter: Developmental Defects

Chapter Section: Miscellaneous Syndromes

TB Case Number: 899

Comments: Axial Mesodermal Dysplasia, Aural atresia

Gender: Female

Otologic Diagnosis:

Axial mesodermal dysplasia with multiple anomalies including:

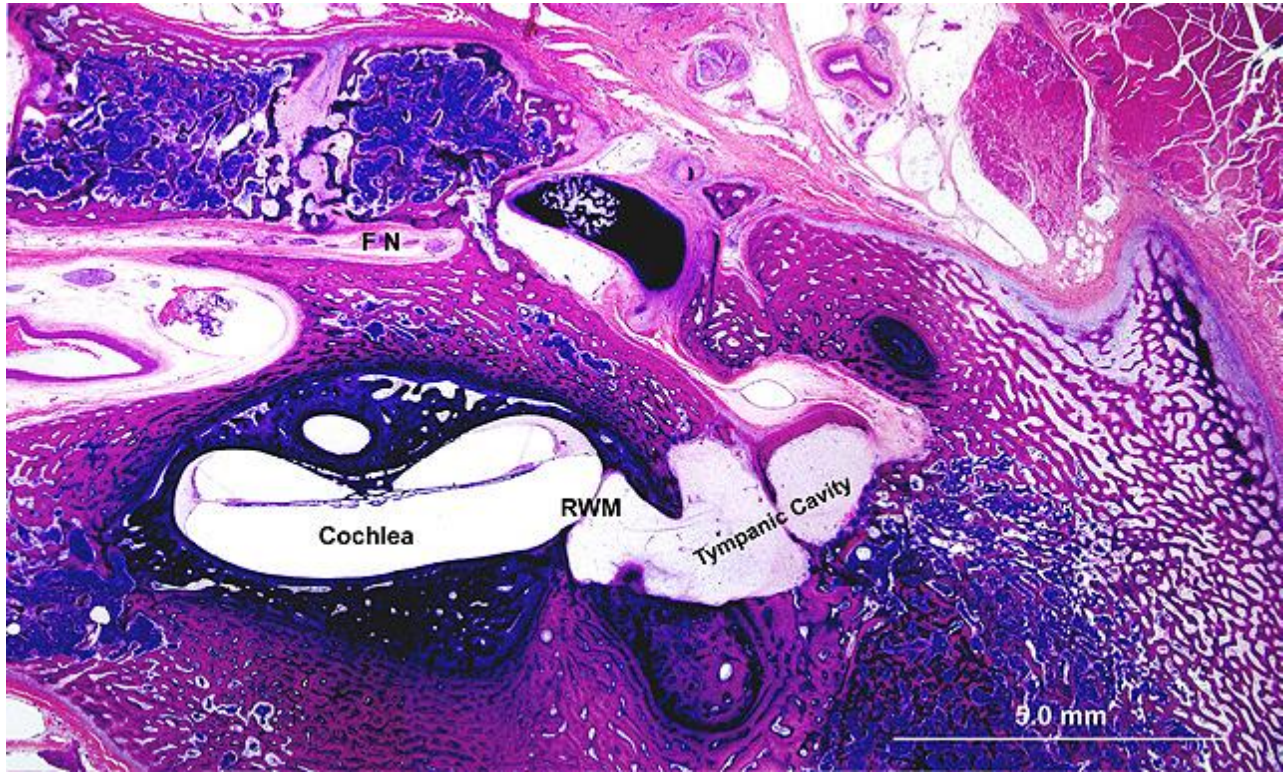
a) Microtia and congenital aural atresia

b) Tympanic cavity, hypoplastic and nonpneumatized

c) Mastoid antrum, non pneumatized

d) Ossicles, dysmorphic

e) Tensor tympani muscle, anomalous location



Axial Mesodermal Dysplasia, R-241

Title: Axial Mesodermal Dysplasia, R-241

Chapter: Developmental Defects

Chapter Section: Miscellaneous Syndromes

TB Case Number: 899

Comments: Axial mesodermal dysplasia, Aural atresia

Gender: Female

Otologic Diagnosis:

Axial mesodermal dysplasia with multiple anomalies including:

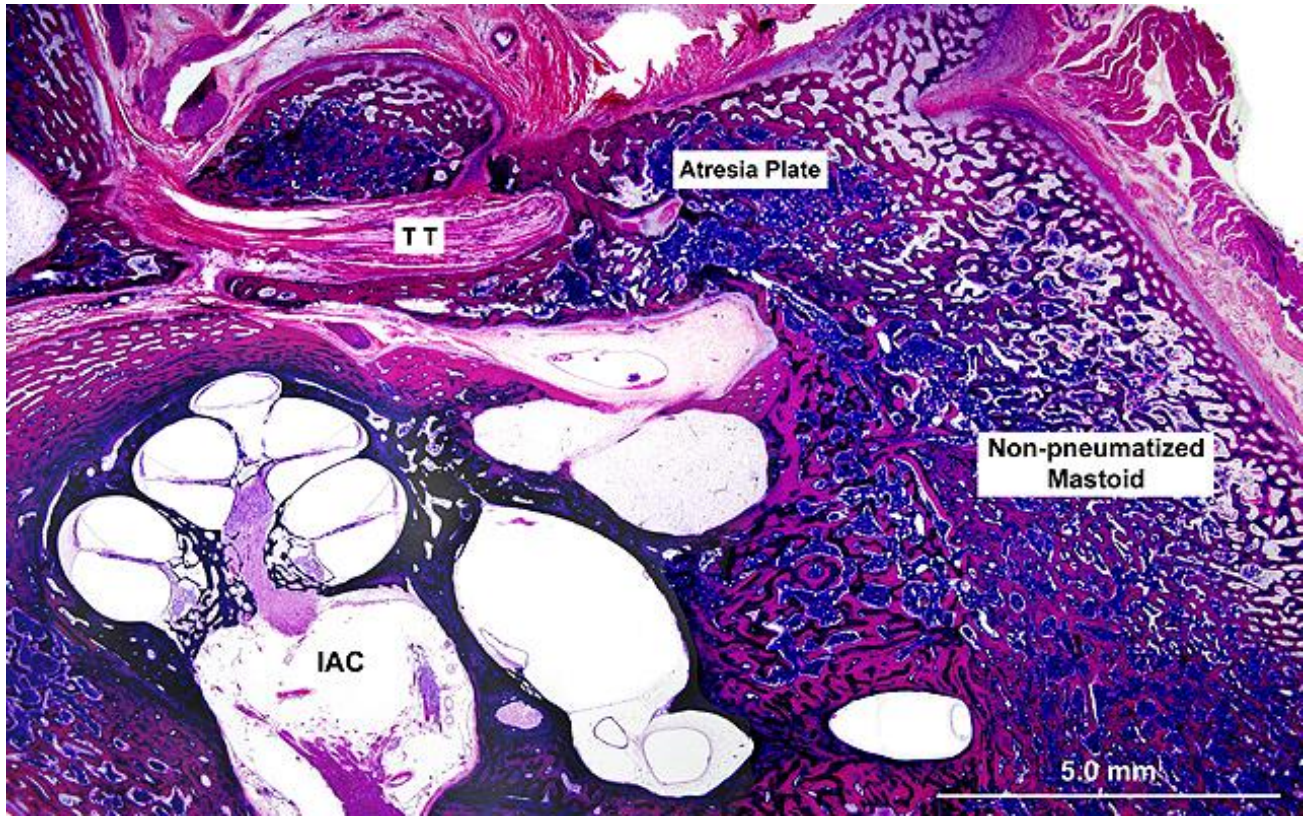
a) Microtia and congenital aural atresia

b) Tympanic cavity, hypoplastic and nonpneumatized

c) Mastoid antrum, non pneumatized

d) Ossicles, dysmorphic

e) Tensor tympani muscle, anomalous location



Cockayne Syndrome, R-191

Title: Cockayne Syndrome, R-191

Chapter: Developmental Defects

Chapter Section: Miscellaneous Syndromes

TB Case Number: 914

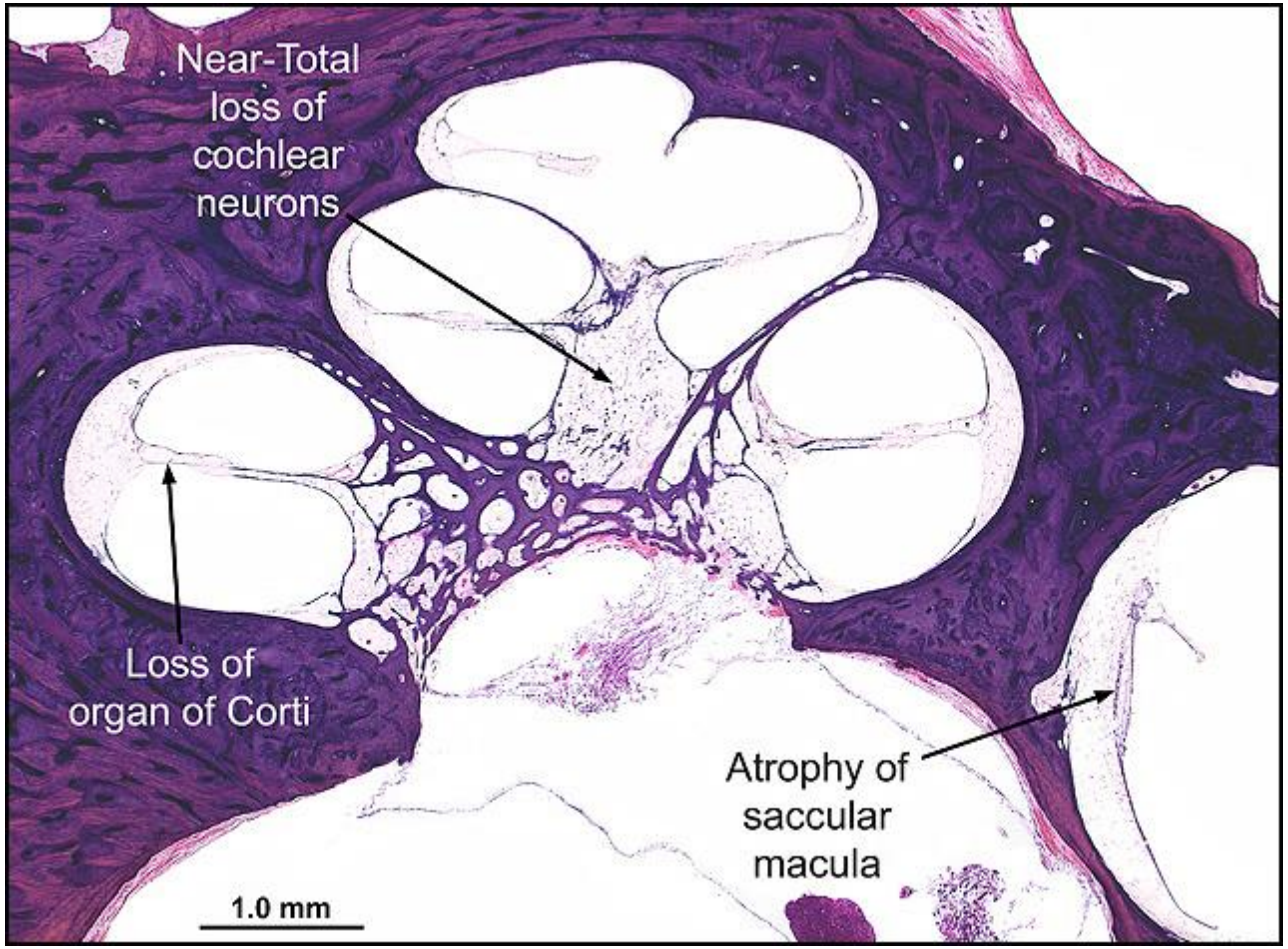
Comments: Cockayne syndrome- showing loss of organ of Corti, stria, spiral ganglion cells, collapse of Reissners and atrophy of saccular macula

Gender: Male

Age (yrs.): 32

Otologic Diagnosis:

1. Cockayne's syndrome with bilateral, profound sensorineural hearing loss
2. Severe degeneration of cochlear and vestibular labyrinths, bilateral
3. Minor developmental anomalies of middle and inner ears, bilateral



Charcot Marie Tooth Syndrome, R-110

Title: Charcot Marie Tooth Syndrome, R-110

Chapter: Developmental Defects

Chapter Section: Miscellaneous Syndromes

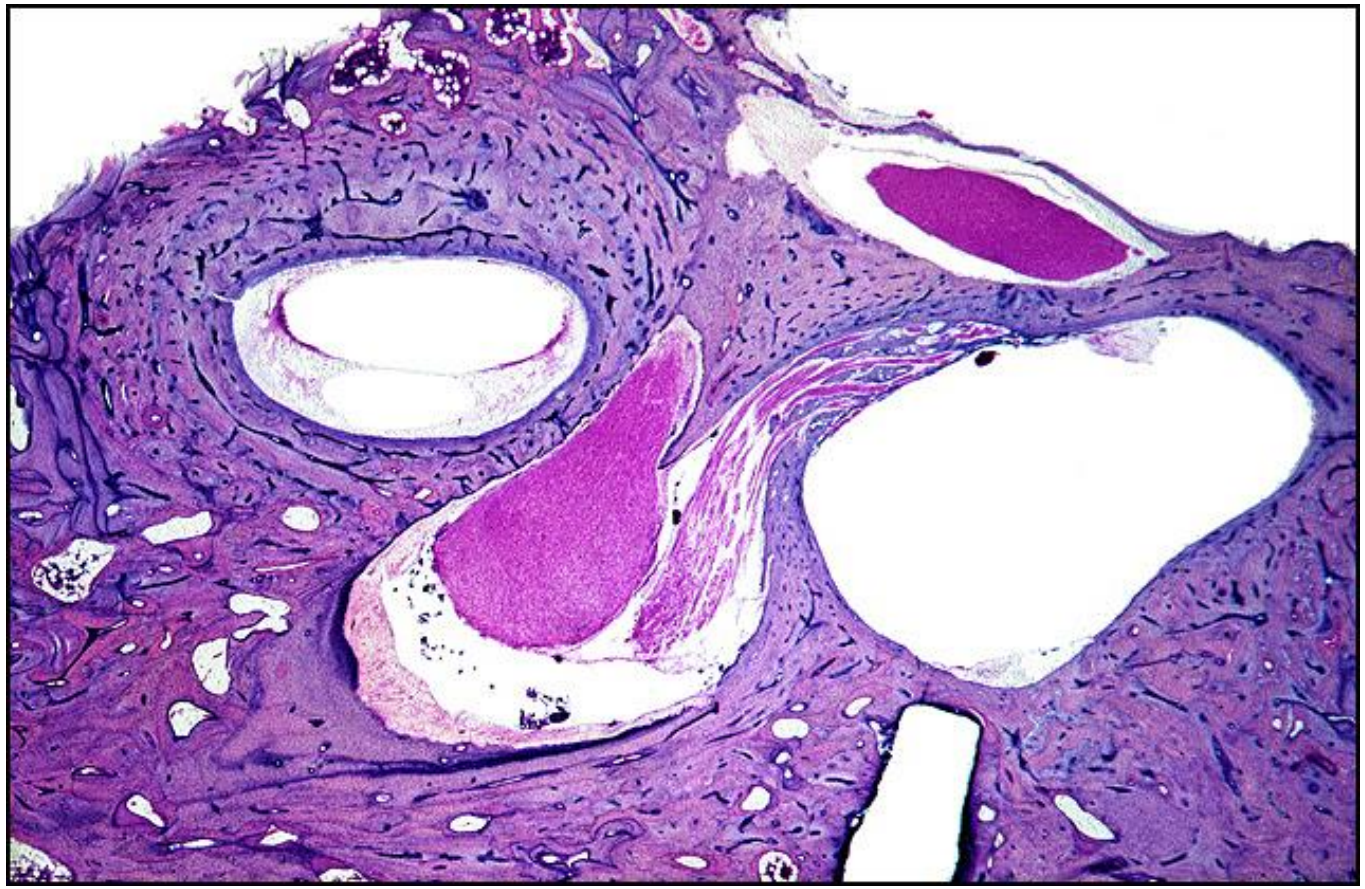
TB Case Number: 691

Gender: Male

Age (yrs.): 74

Otologic Diagnosis:

1. Charcot-Marie-Tooth syndrome
2. Sensorineural hearing loss, severe, bilateral (neuronal and strial atrophy)
3. Hypertrophy of the facial nerves, bilateral



Kleeblattschadel, L-181

Title: Kleeblattschadel, L-181

Chapter: Developmental Defects

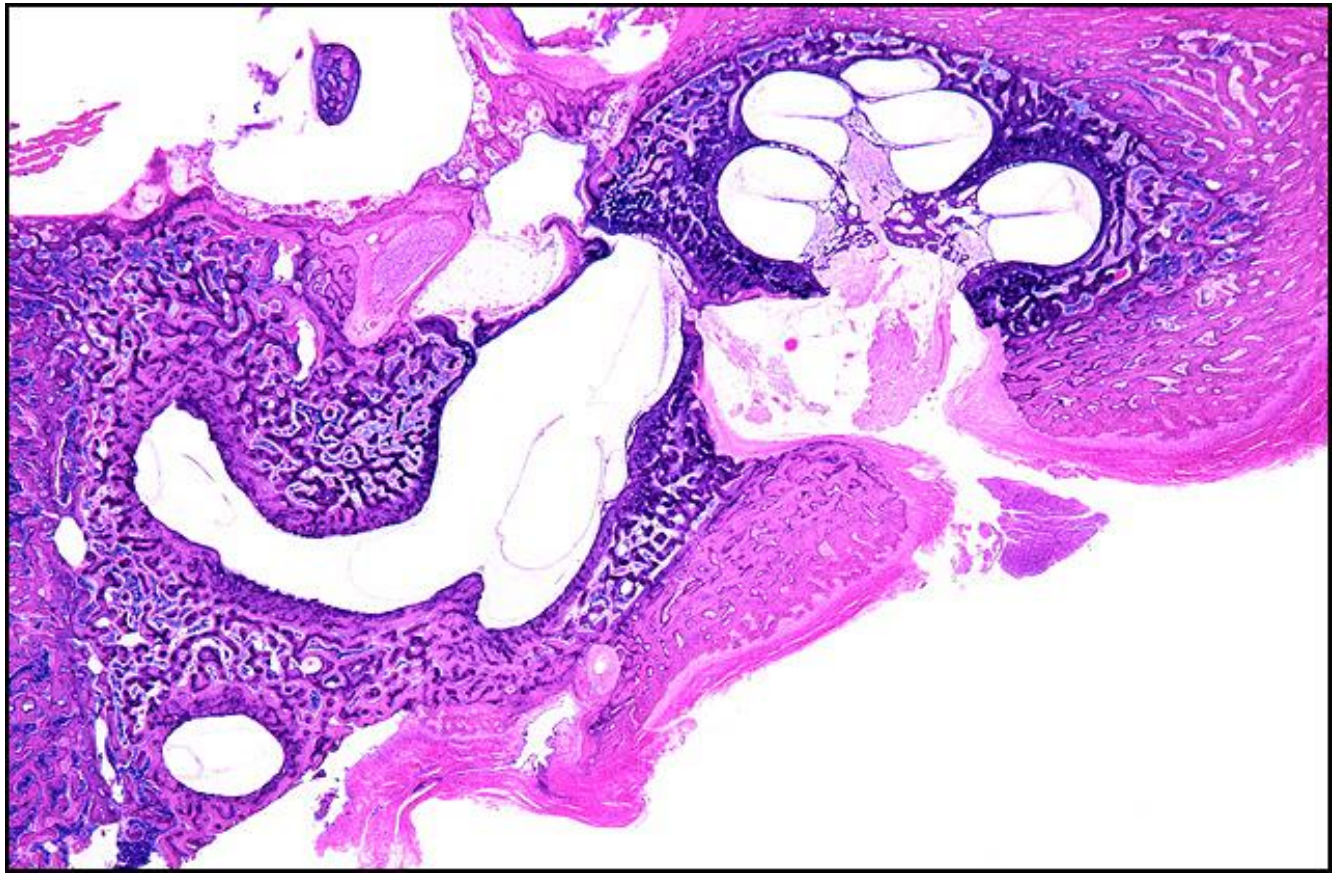
Chapter Section: Miscellaneous Syndromes

TB Case Number: 161

Gender: Female

Otologic Diagnosis:

1. Thanatophoric skeletal dysplasia with cloverleaf skull deformity (Kleeblattschädel)
2. Hydrocephalus, severe
3. Endolymphatic hydrops, bilateral, possibly secondary to hydrocephalus, bilateral
4. Stapes, fetal configuration, bilateral



Kleeblattschadel, R-181

Title: Kleeblattschadel, R-181

Chapter: Developmental Defects

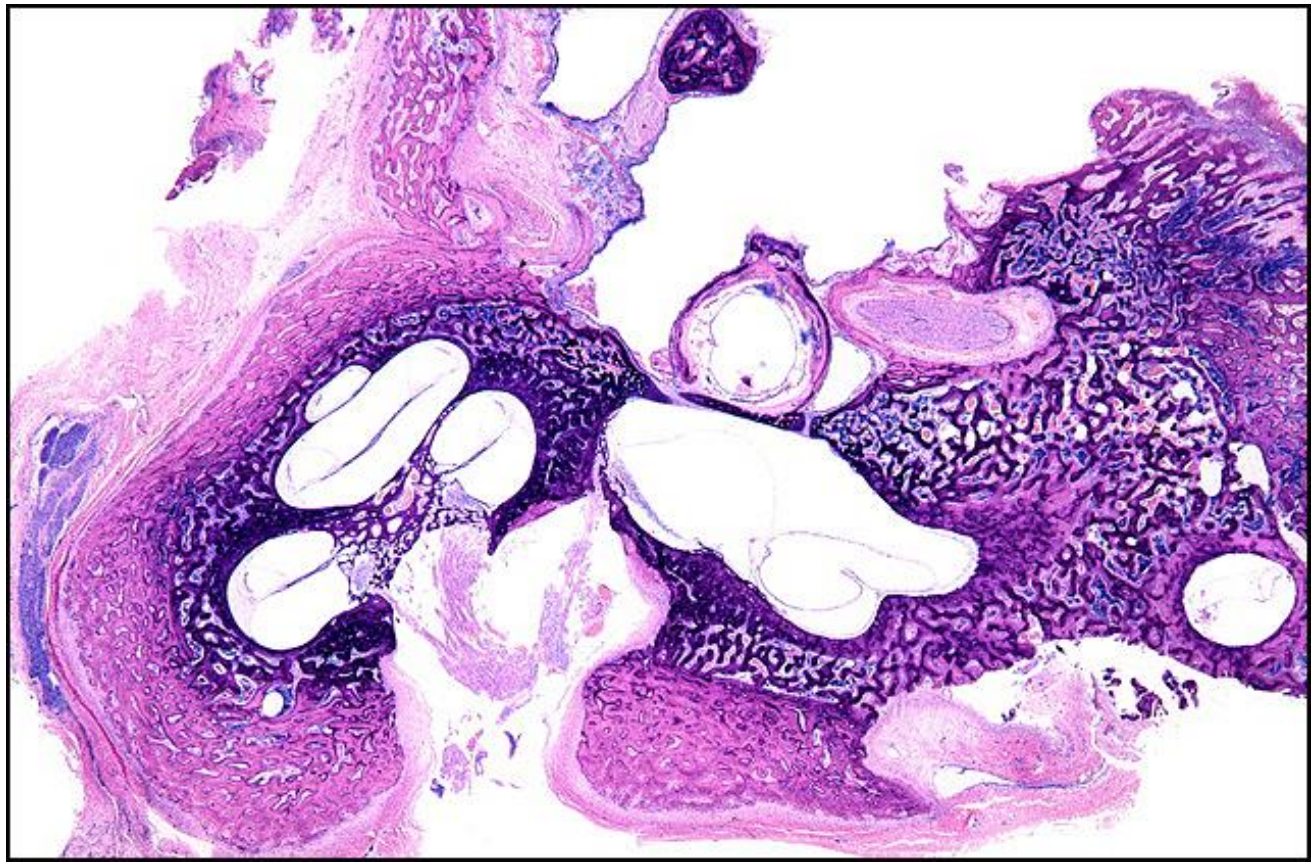
Chapter Section: Miscellaneous Syndromes

TB Case Number: 161

Gender: Female

Otologic Diagnosis:

1. Thanatophoric skeletal dysplasia with cloverleaf skull deformity (Kleeblattschädel)
2. Hydrocephalus, severe
3. Endolymphatic hydrops, bilateral, possibly secondary to hydrocephalus, bilateral
4. Stapes, fetal configuration, bilateral



Cockayne's Syndrome, R-221

Title: Cockayne's Syndrome, R-221

Chapter: Developmental Defects

Chapter Section: Miscellaneous Syndromes

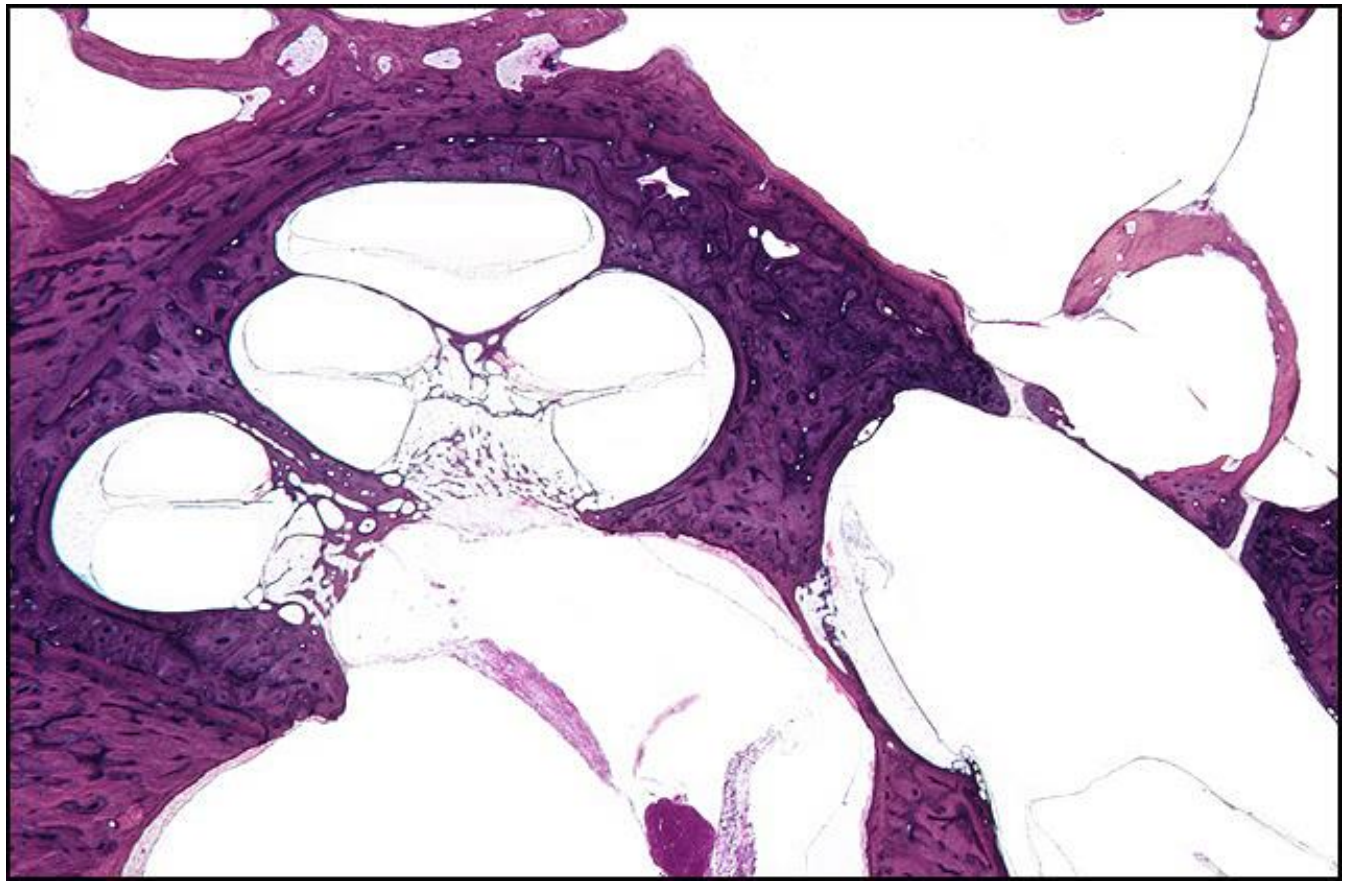
TB Case Number: 914

Gender: Male

Age (yrs.): 32

Otologic Diagnosis:

1. Cockayne's syndrome with bilateral, profound sensorineural hearing loss
2. Severe degeneration of cochlear and vestibular labyrinths, bilateral
3. Minor developmental anomalies of middle and inner ears, bilateral



Cockayne's Syndrome, R-101

Title: Cockayne's syndrome, R-101

Chapter: Developmental Defects

Chapter Section: Miscellaneous Syndromes

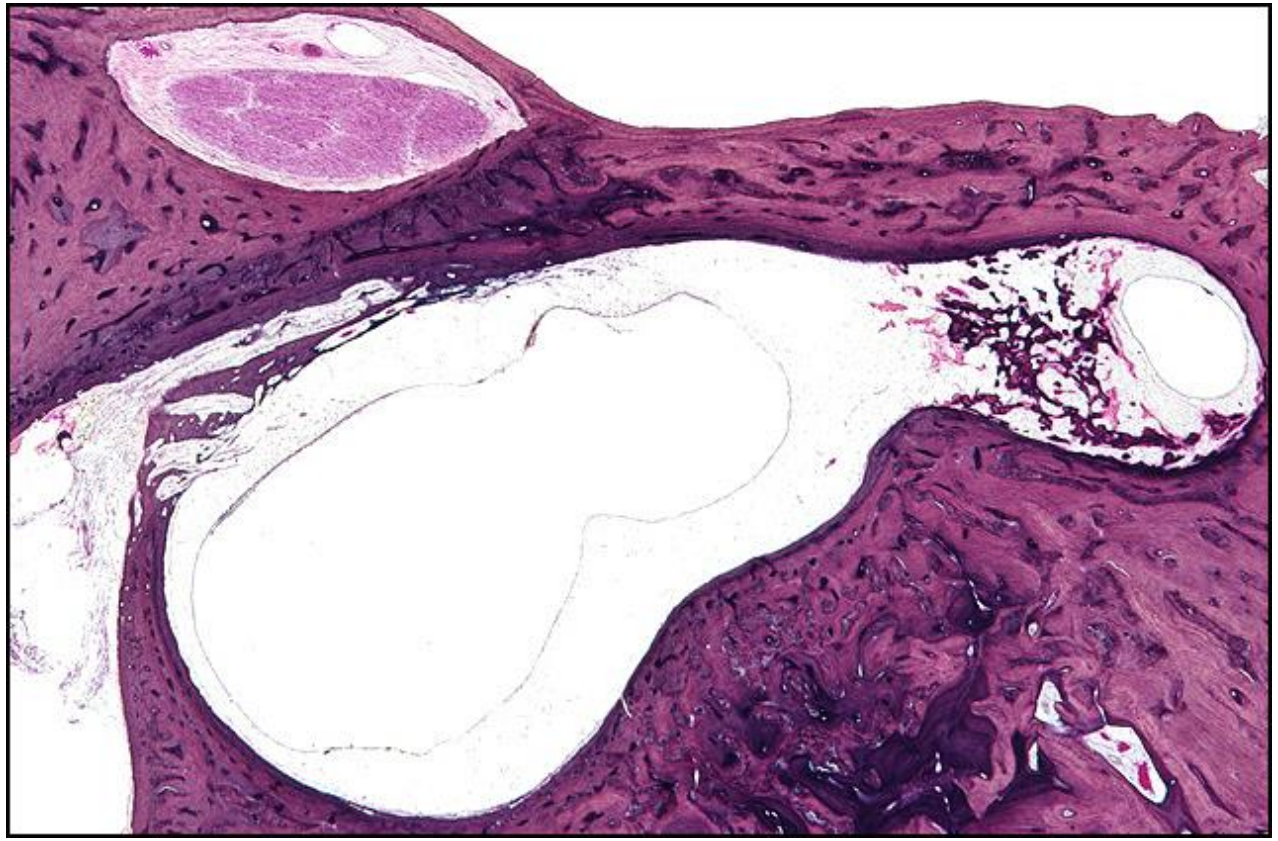
TB Case Number: 914

Gender: Male

Age (yrs.): 32

Otologic Diagnosis:

1. Cockayne's syndrome with bilateral, profound sensorineural hearing loss
2. Severe degeneration of cochlear and vestibular labyrinths, bilateral
3. Minor developmental anomalies of middle and inner ears, bilateral



Cockayne's Syndrome, R-321

Title: Cockayne's Syndrome, R-321

Chapter: Developmental Defects

Chapter Section: Miscellaneous Syndromes

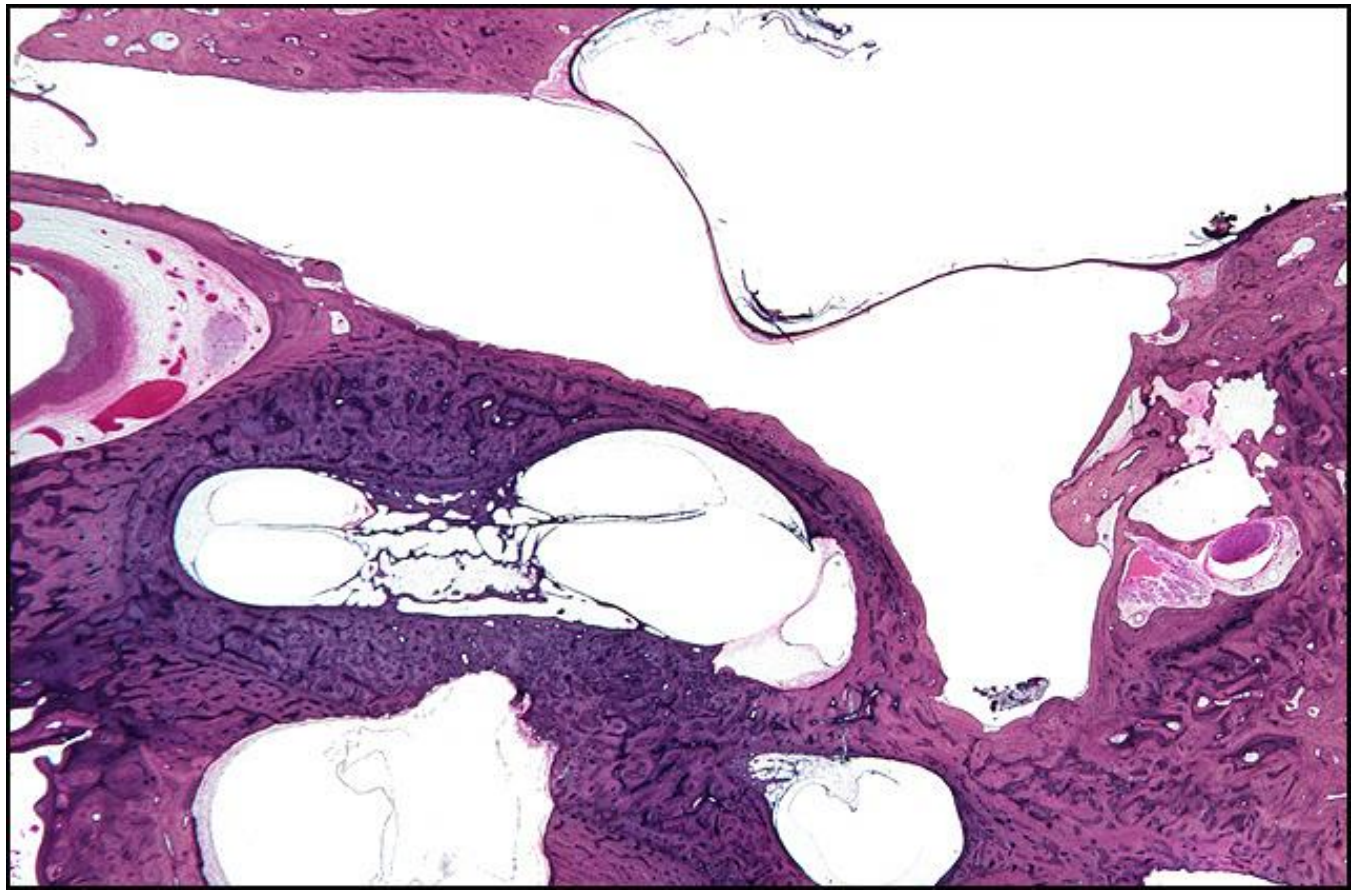
TB Case Number: 914

Gender: Male

Age (yrs.): 32

Otologic Diagnosis:

1. Cockayne's syndrome with bilateral, profound sensorineural hearing loss
2. Severe degeneration of cochlear and vestibular labyrinths, bilateral
3. Minor developmental anomalies of middle and inner ears, bilateral



Maternal Rubella, L-191

Title: Maternal Rubella, L-191

Chapter: Developmental Defects

Chapter Section: Noxious Prenatal Influences / Maternal Rubella

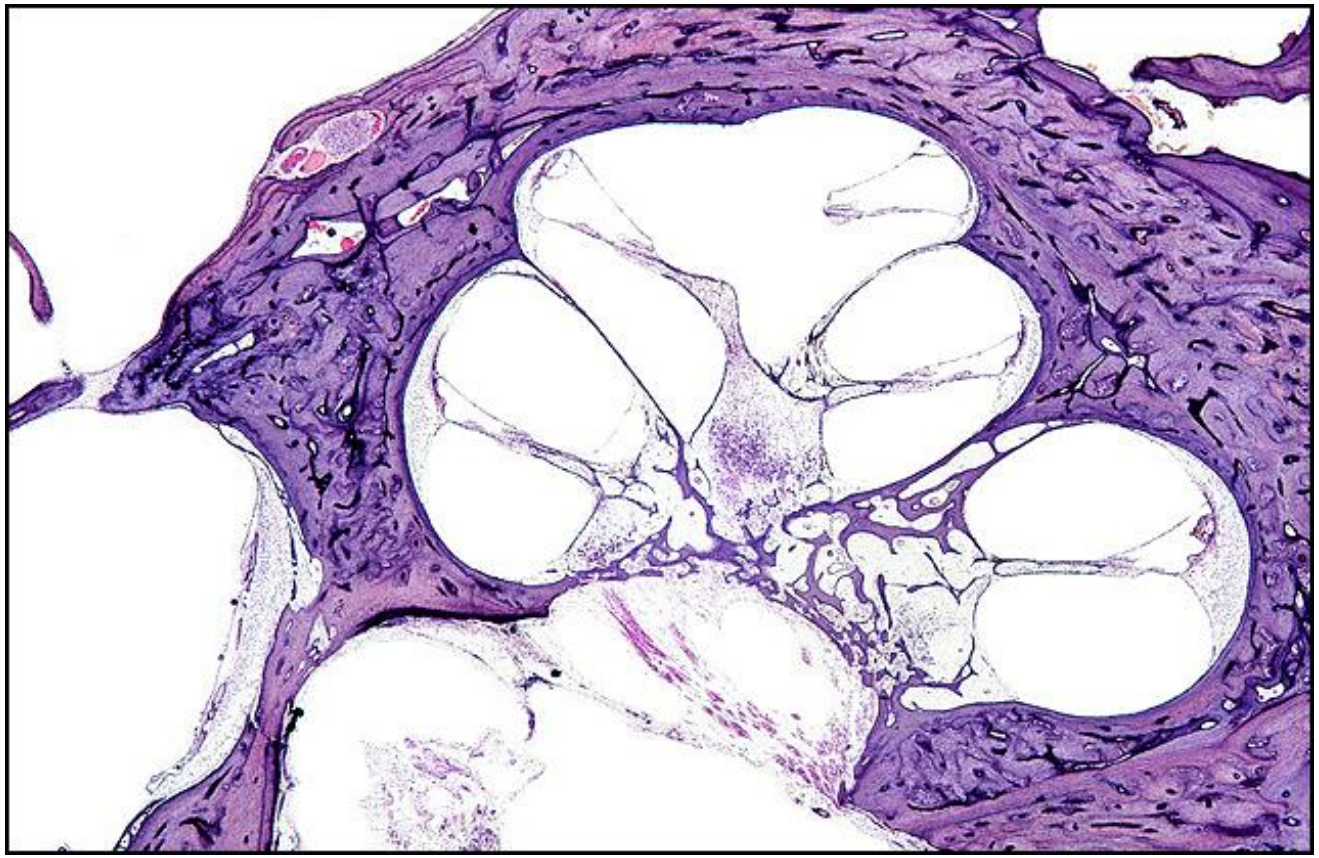
TB Case Number: 347

Gender: Female

Age (yrs.): 58

Otologic Diagnosis:

1. Cochleosaccular degeneration, severe, bilateral
2. Normal pars superior (utricle and canals), bilateral
3. Retrograde cochlear neuronal degeneration, bilateral, secondary to #1
4. Deaf-mutism caused by maternal rubella, diagnosis by exclusion on basis of pathology



Maternal Rubella, R-71

Title: Maternal Rubella, R-71

Chapter: Developmental Defects

Chapter Section: Noxious Prenatal Influences / Maternal Rubella

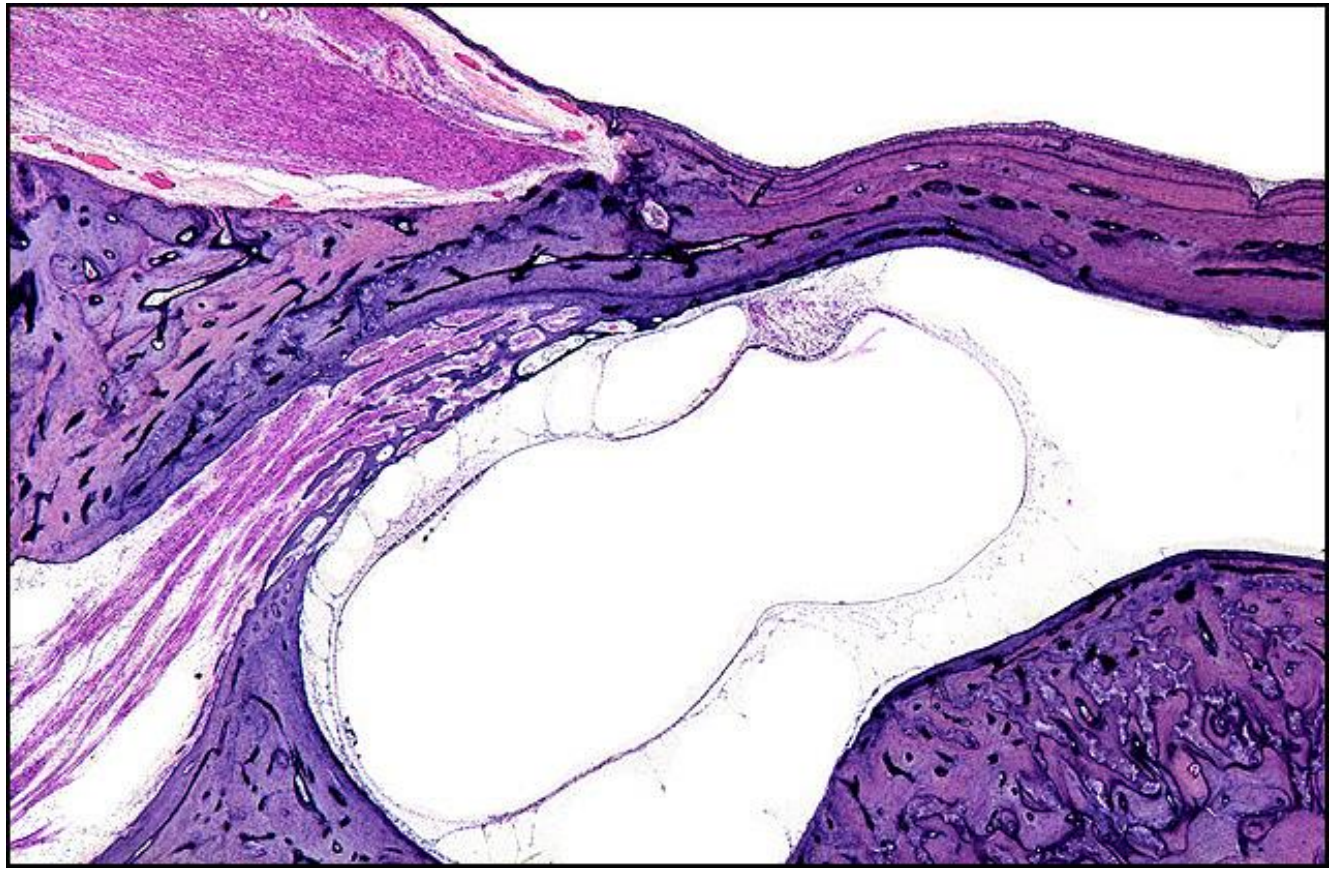
TB Case Number: 347

Gender: Female

Age (yrs.): 58

Otologic Diagnosis:

1. Cochleosaccular degeneration, severe, bilateral
2. Normal pars superior (utricle and canals), bilateral
3. Retrograde cochlear neuronal degeneration, bilateral, secondary to #1
4. Deaf-mutism caused by maternal rubella, diagnosis by exclusion on basis of pathology



Maternal Rubella, R-196

Title: Maternal Rubella, R-196

Chapter: Developmental Defects

Chapter Section: Noxious Prenatal Influences / Maternal Rubella

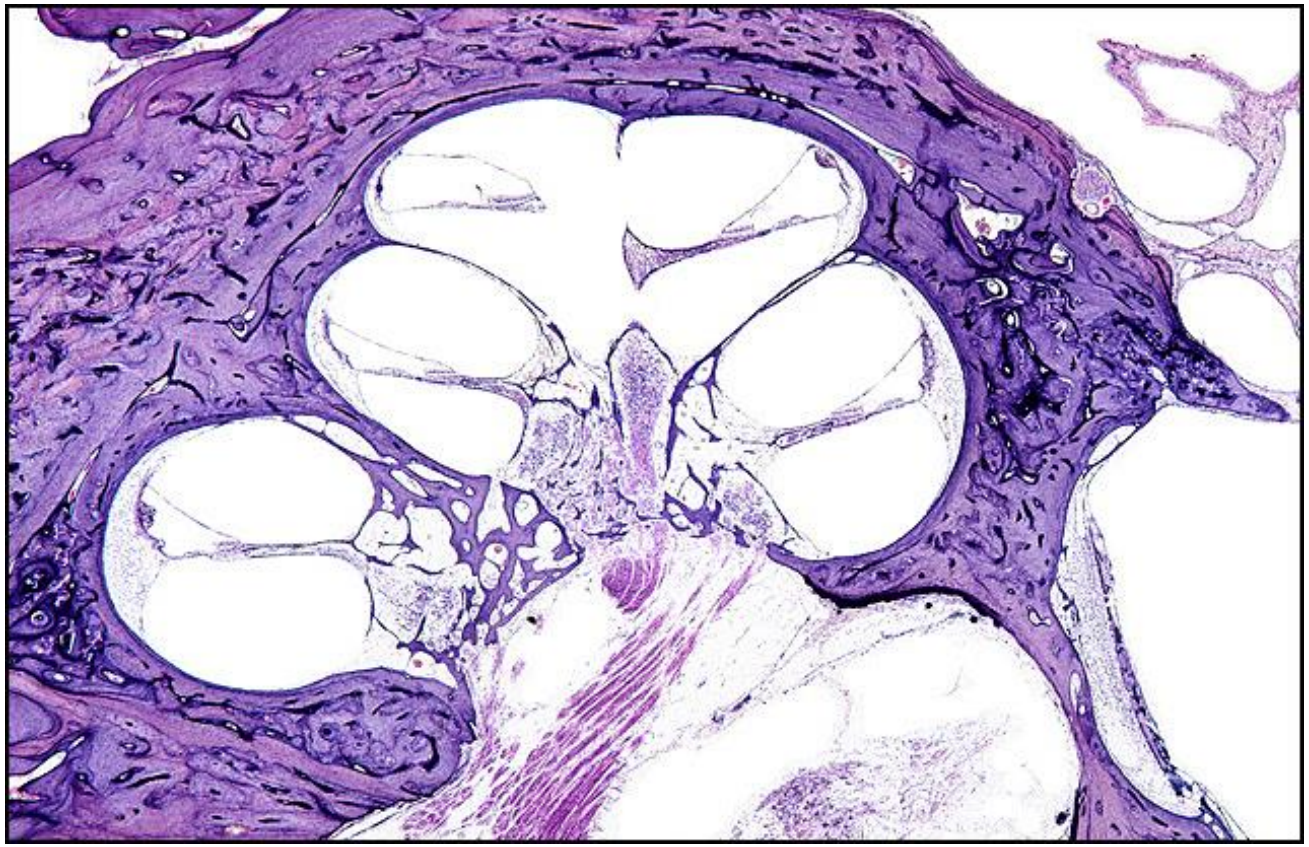
TB Case Number: 347

Gender: Female

Age (yrs.): 58

Otologic Diagnosis:

1. Cochleosaccular degeneration, severe, bilateral
2. Normal pars superior (utricle and canals), bilateral
3. Retrograde cochlear neuronal degeneration, bilateral, secondary to #1
4. Deaf-mutism caused by maternal rubella, diagnosis by exclusion on basis pathology



Axial Mesodermal Dysplasia, R-351

Title: Axial Mesodermal Dysplasia, R-351

Chapter: Developmental Defects

Chapter Section: Variform Anomalies External and Middle Ear / Congenital Aural Atresia

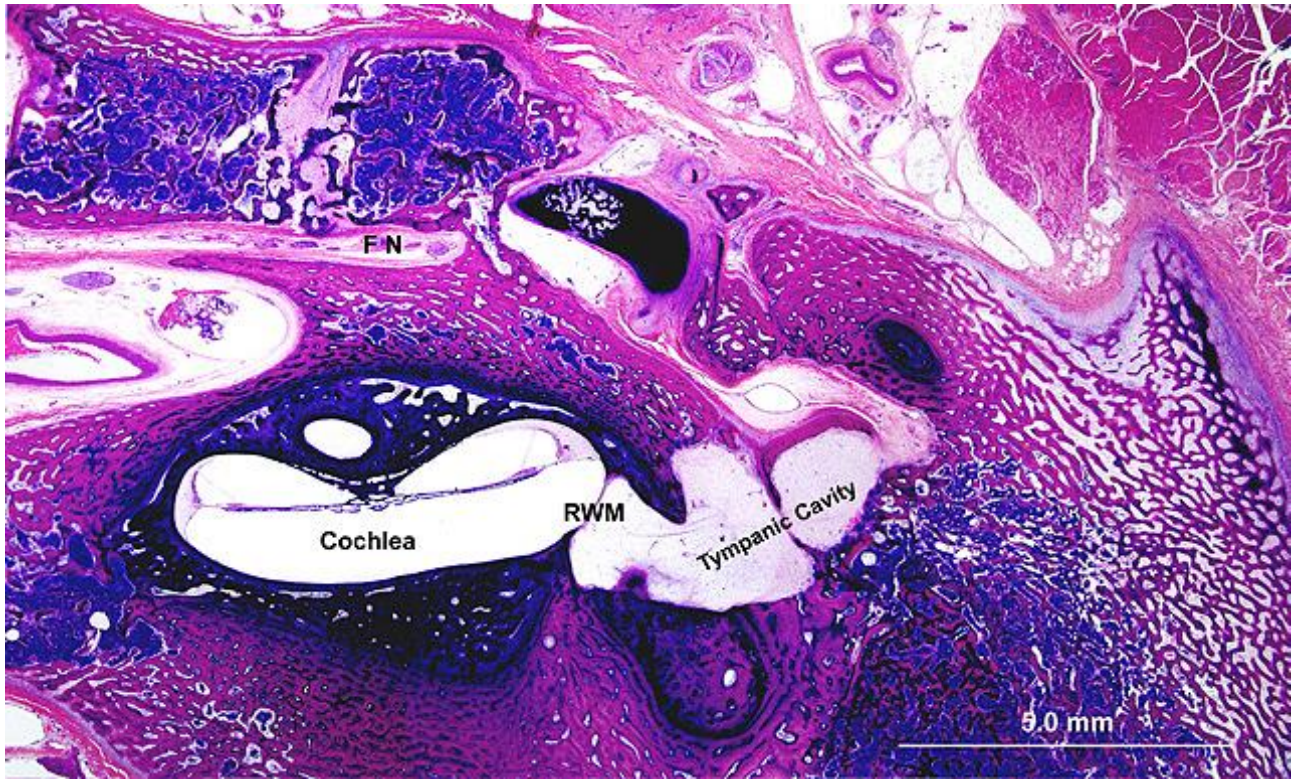
TB Case Number: 899

Comments: Axial mesodermal dysplasia, Aural atresia

Gender: Female

Otologic Diagnosis: Axial mesodermal dysplasia with multiple anomalies including:

1. Microtia and congenital aural atresia
2. Tympanic cavity, hypoplastic and nonpneumatized
3. Mastoid antrum, non pneumatized
4. Ossicles, dysmorphic
5. Tensor tympani muscle, anomalous location



Axial Mesodermal Dysplasia, R-241

Title: Axial Mesodermal Dysplasia, R-241

Chapter: Developmental Defects

Chapter Section: Variform Anomalies External and Middle Ear / Congenital Aural Atresia

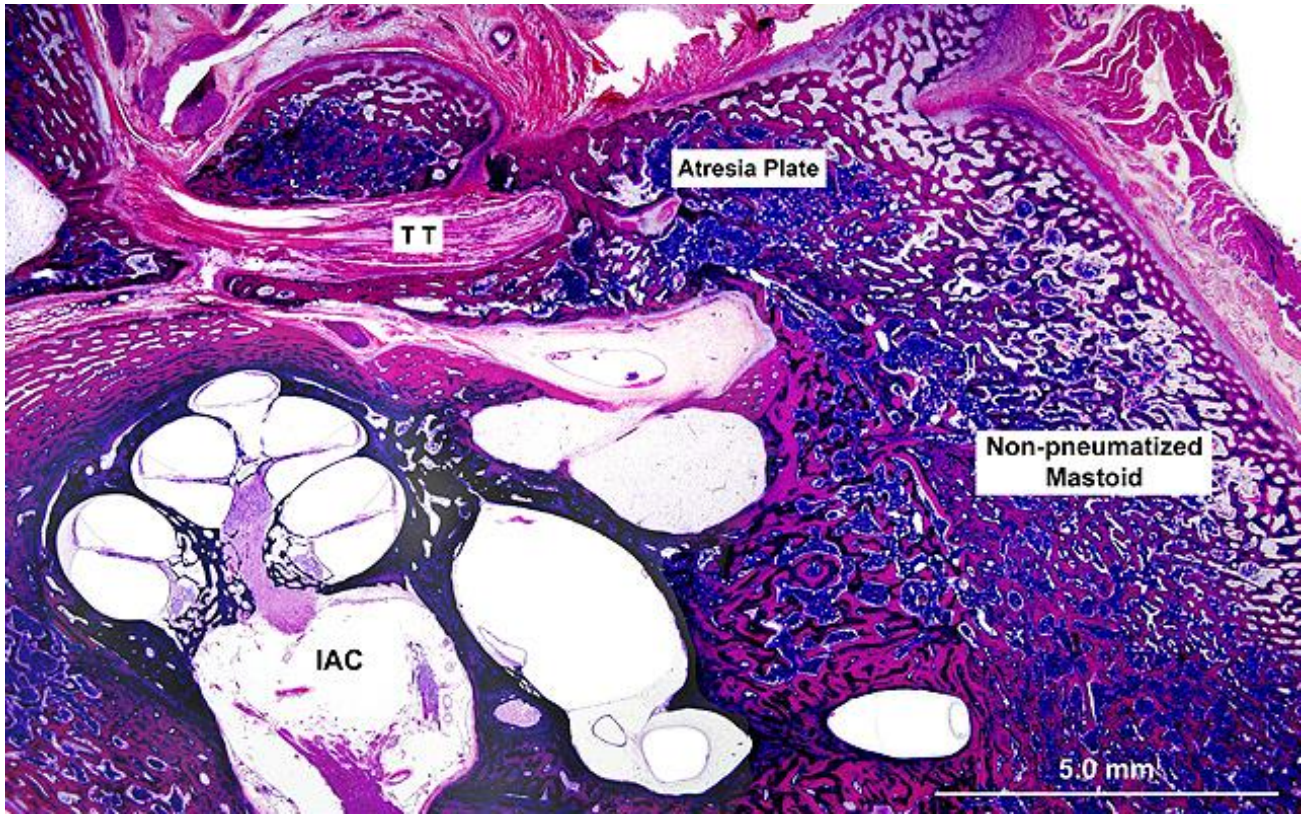
TB Case Number: 899

Comments: Axial mesodermal dysplasia, Aural atresia

Gender: Female

Otologic Diagnosis: Axial mesodermal dysplasia with multiple anomalies including:

1. Microtia and congenital aural atresia
2. Tympanic cavity, hypoplastic and non-pneumatized
3. Mastoid antrum, non pneumatized
4. Ossicles, dysmorphic
5. Tensor tympani muscle, anomalous location



Congenital Aural Atresia, R-261

Title: Congenital Aural Atresia, R-261

Chapter: Developmental Defects

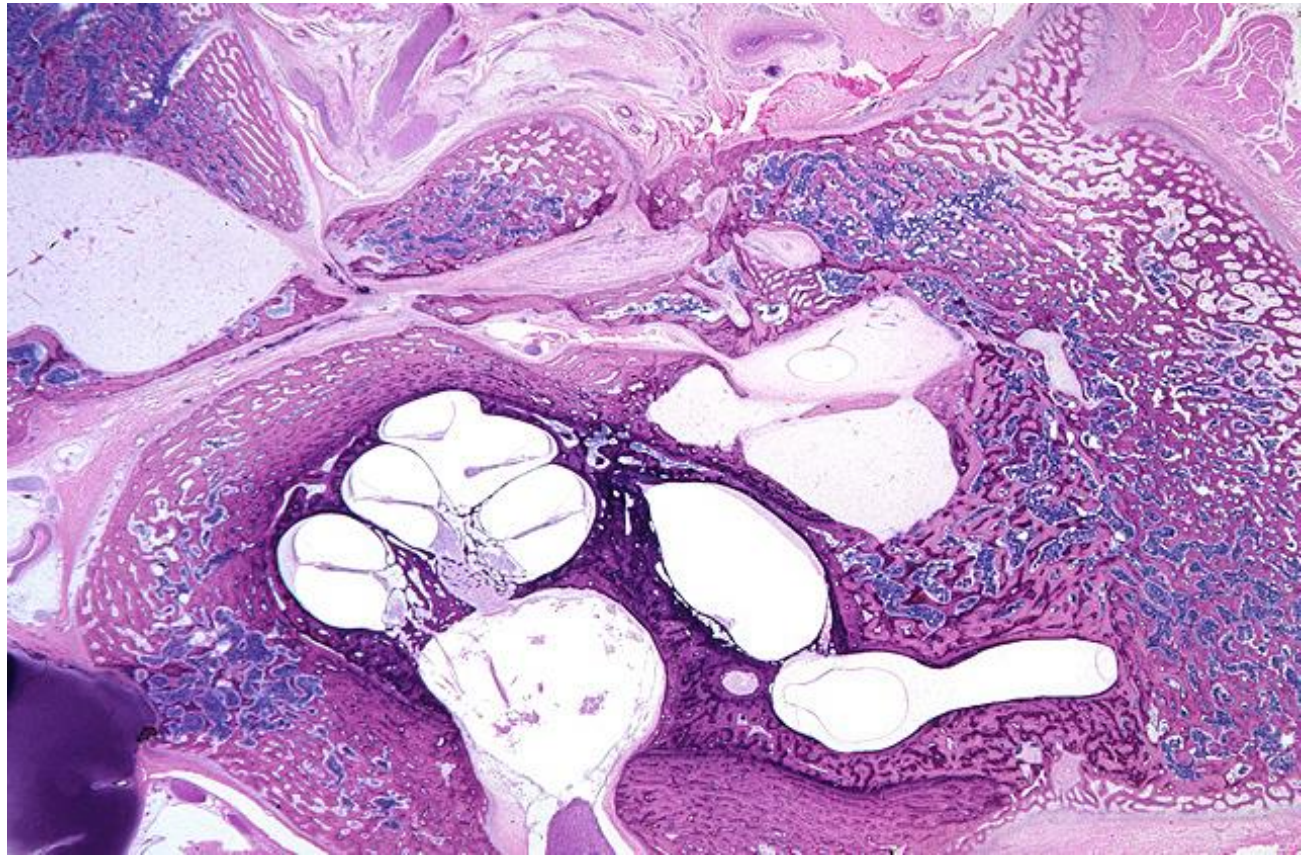
Chapter Section: Variform Anomalies External and Middle Ear / Congenital Aural Atresia

TB Case Number: 899

Gender: Female

Otologic Diagnosis: Axial mesodermal dysplasia with multiple anomalies including:

1. Microtia and congenital aural atresia
2. Tympanic cavity, hypoplastic and non-pneumatized
3. Mastoid antrum, non pneumatized
4. Ossicles, dysmorphic
5. Tensor tympani muscle, anomalous location



High Jugular Bulb, R-411

Title: High Jugular Bulb, R-411

Chapter: Developmental Defects

Chapter Section: Variform Anomalies External and Middle Ear / Miscellaneous

TB Case Number: 972

Comments: high jugular bulb dehiscent into the hypotympanum in 92 yr. old female, no symptoms

Gender: Female

Age (yrs.): 93

Otologic Diagnosis:

Clinical:

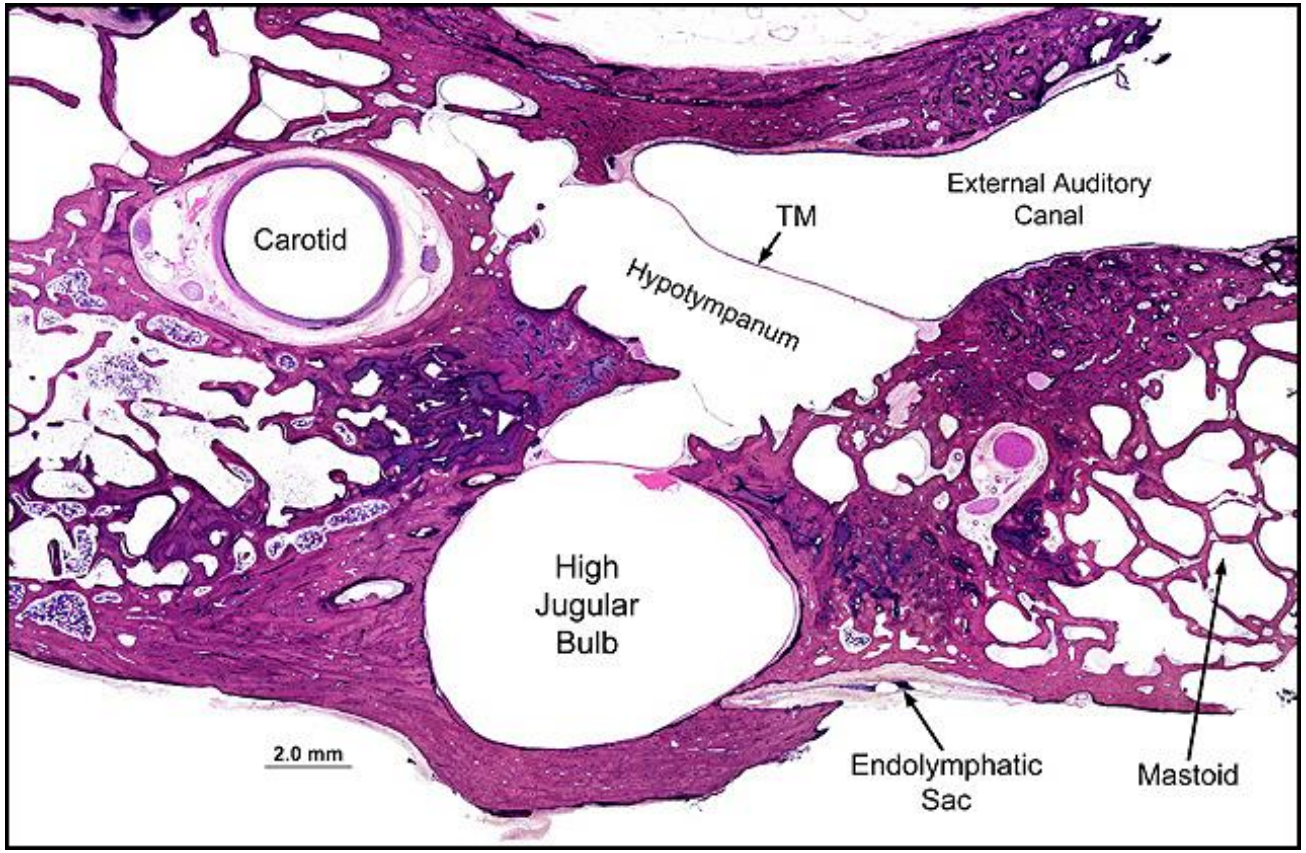
Sensorineural hearing loss, down-sloping, consistent with presbycusis

Histopathologic:

Cochlear pathology, consistent with mixed presbycusis:

a) Complete atrophy of organ of Corti, basal turn

b) Partial loss of outer hair cells



Malleus Fixation, L-11

Title: Malleus Fixation, L-11

Chapter: Developmental Defects

Chapter Section: Variform Anomalies External and Middle Ear / Ossicular Anomalies

TB Case Number: 282

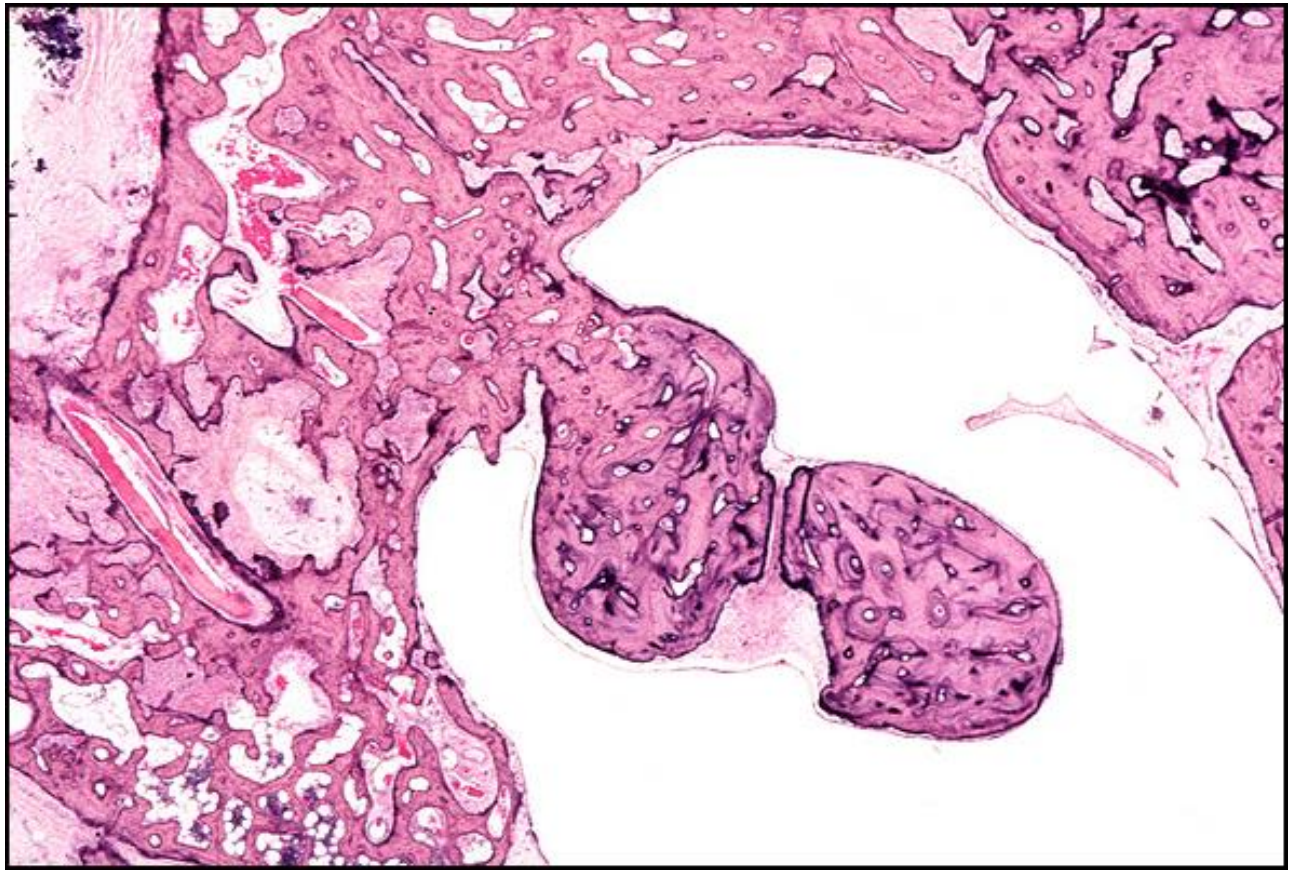
Comments: Malleus fixation

Gender: Female

Age (yrs.): 89

Otologic Diagnosis:

1. Sensorineural hearing loss, severe, flat audiogram, symmetrical, bilateral
2. Presbycusis, indeterminate type
3. Malleus, fixation, epitympanum, congenital anomaly, presumptive, left
4. Exostoses, small, external auditory canals, bilateral



Incus Deformity, R-161

Title: Incus Deformity, R-161

Chapter: Developmental Defects

Chapter Section: Variform Anomalies External and Middle Ear / Ossicular Anomalies

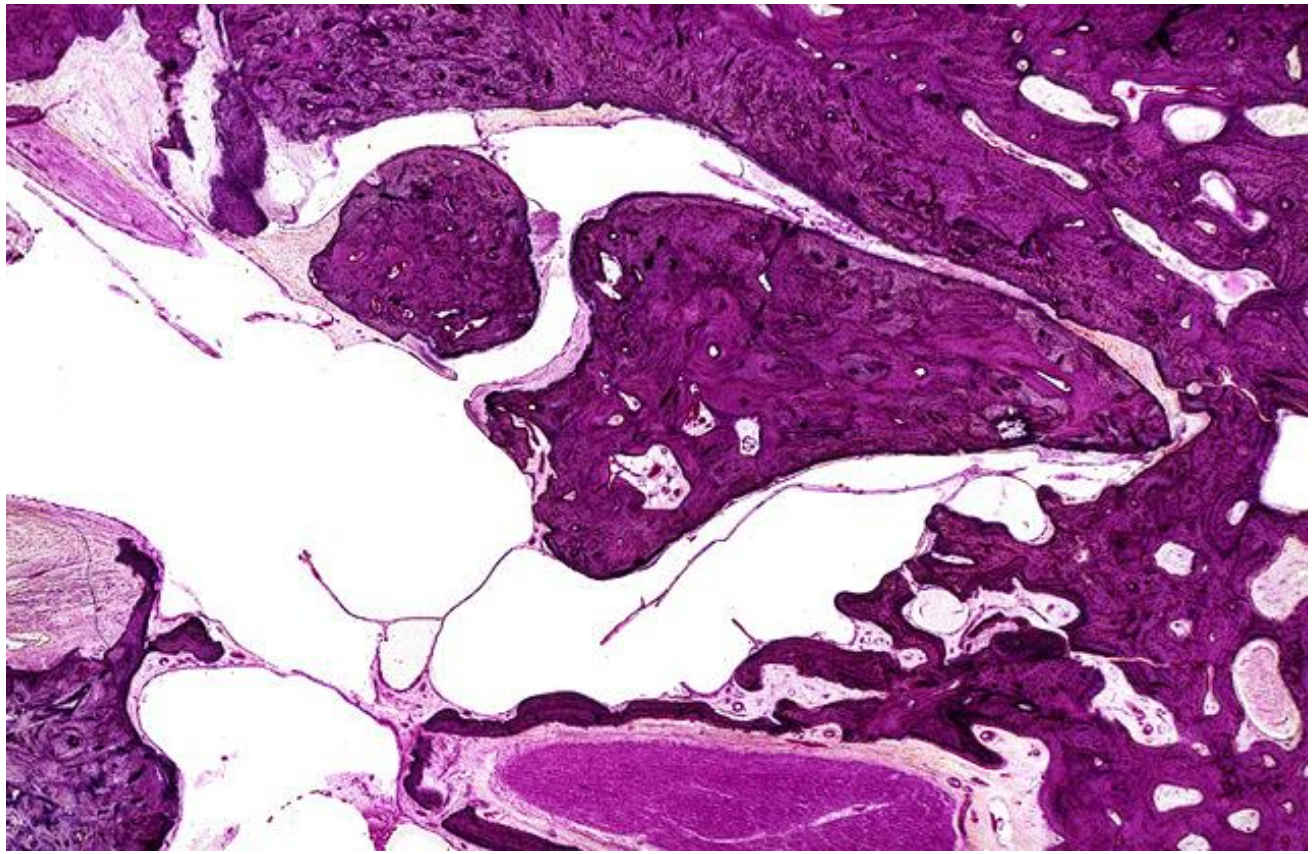
TB Case Number: 121

Gender: Female

Age (yrs.): 64

Otologic Diagnosis:

1. Jakob-Creutzfeldt disease
2. Membranous labyrinth, normal, bilateral
3. Bony labyrinth, normal, bilateral
4. Malleus fixation, epitympanum, osseous, left
5. Incus fixation, mild, right
6. Artifact, postmortem autolysis, bilateral



Incus Deformity, L-61

Title: Incus Deformity, L-61

Chapter: Developmental Defects

Chapter Section: Variform Anomalies External and Middle Ear / Ossicular Anomalies

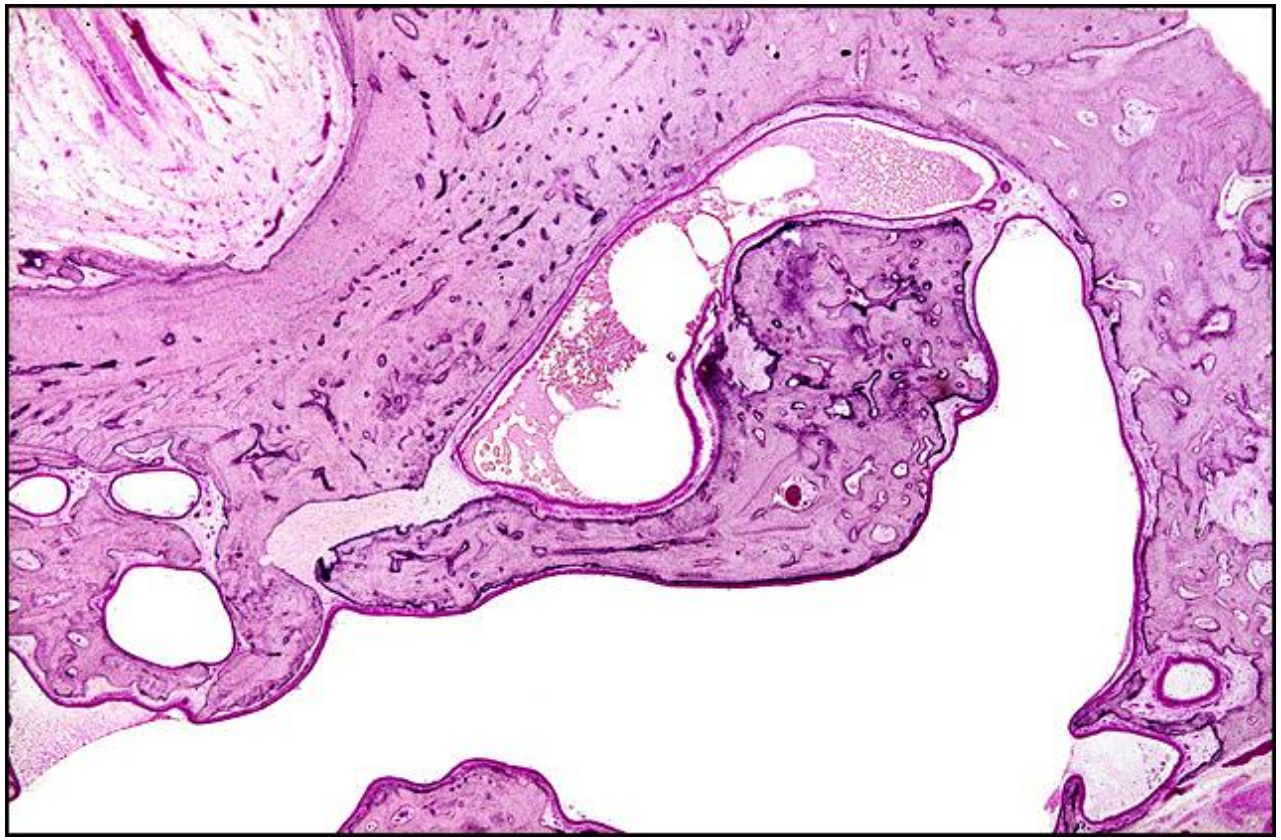
TB Case Number: 407

Gender: Male

Age (yrs.): 19

Otologic Diagnosis:

1. Tympanoplasty, myringoplasty, with obliteration of tympanomeatal angles, bilateral
2. Tympanic membrane, perforation, with migration of mucous membrane into the external auditory canal, right
3. Stapes, fixation, congenital, bilateral
4. Stapes, fracture



Congenital Incus Ankylosis, R-111

Title: Congenital Incus Ankylosis, R-111

Chapter: Developmental Defects

Chapter Section: Variform Anomalies External and Middle Ear / Ossicular Anomalies

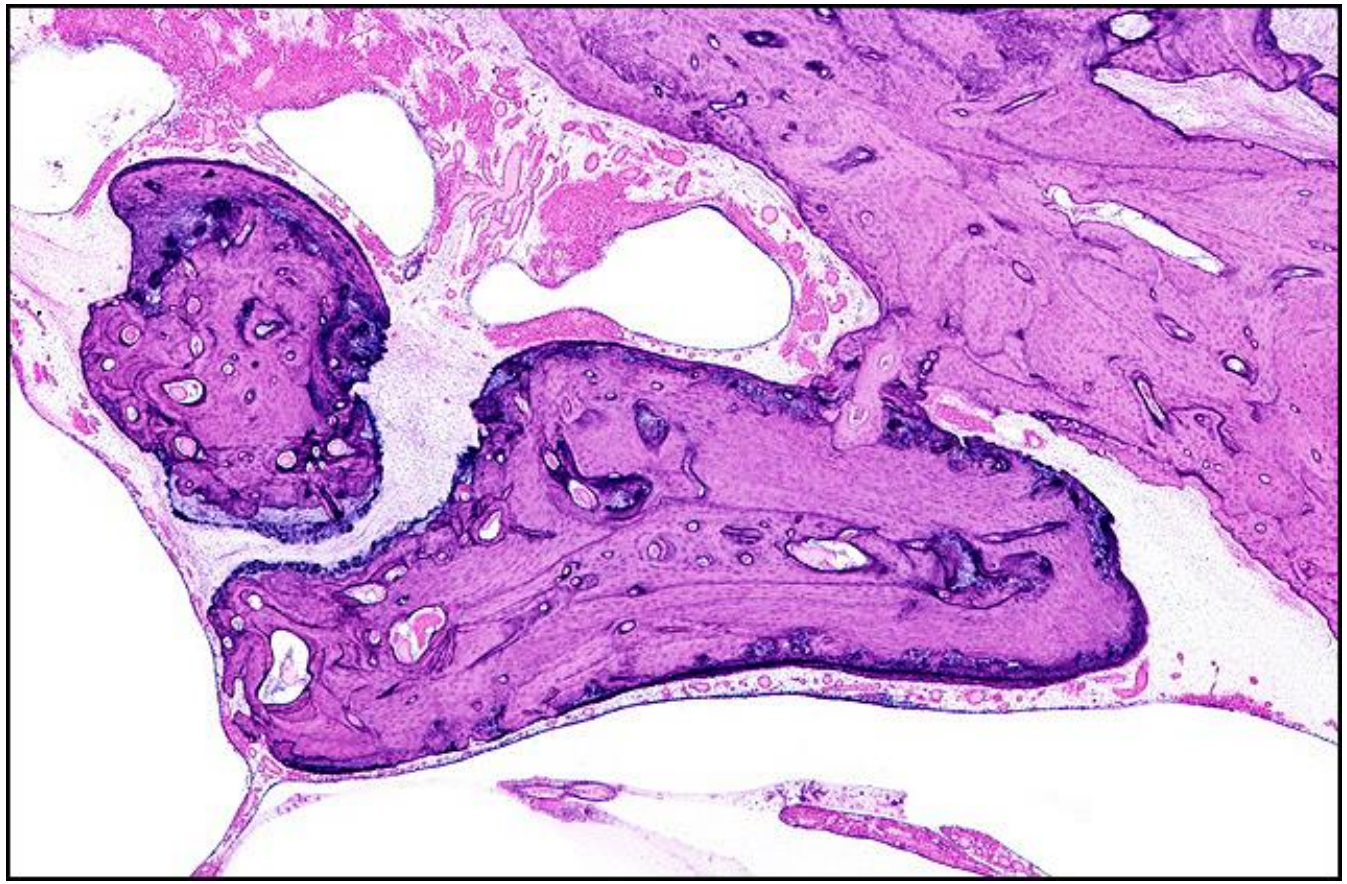
TB Case Number: 904

Gender: Male

Age (yrs.): 6

Otologic Diagnosis:

1. Otitis media, active, chronic
2. Mastoiditis, active, chronic
3. Effusion, middle ear and mastoid, chronic
4. Incus, anomaly, ankylosis
5. Anomaly, cochlea, mild, enlarged Scala tympani in basal and middle turns
6. Saccule, endolymphatic distension



Congenital Incus Ankylosis, R-111

Title: Congenital Incus Ankylosis, R-111

Chapter: Developmental Defects

Chapter Section: Variform Anomalies External and Middle Ear / Ossicular Anomalies

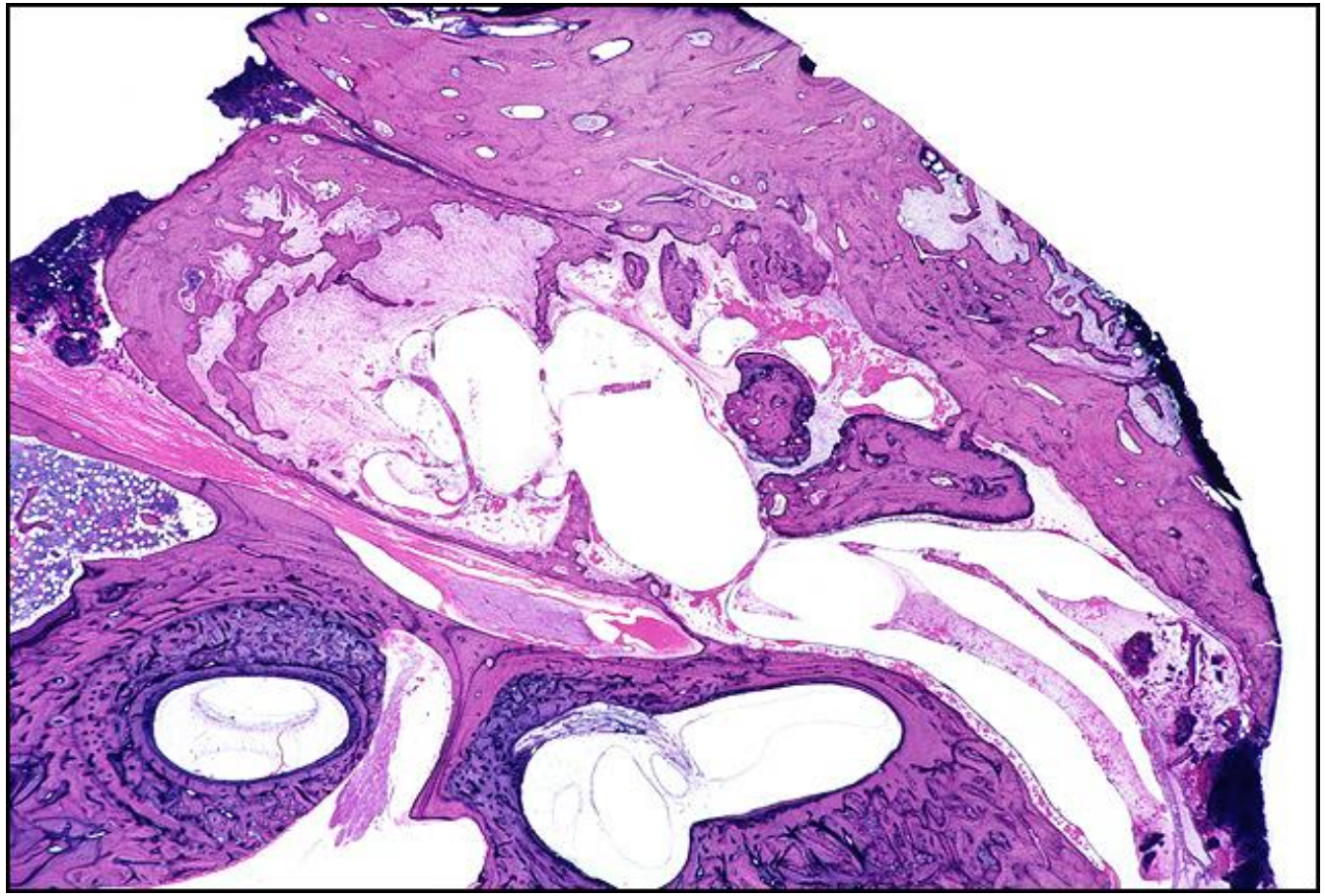
TB Case Number: 904

Gender: Male

Age (yrs.): 6

Otologic Diagnosis:

1. Otitis media, active, chronic
2. Mastoiditis, active, chronic
3. Effusion, middle ear and mastoid, chronic
4. Incus, anomaly, ankylosis
5. Anomaly, cochlea, mild, enlarged Scala tympani in basal and middle turns
6. Saccule, endolymphatic distension



Persistent Stapedial Artery, L-291

Title: Persistent Stapedial Artery, L-291

Chapter: Developmental Defects

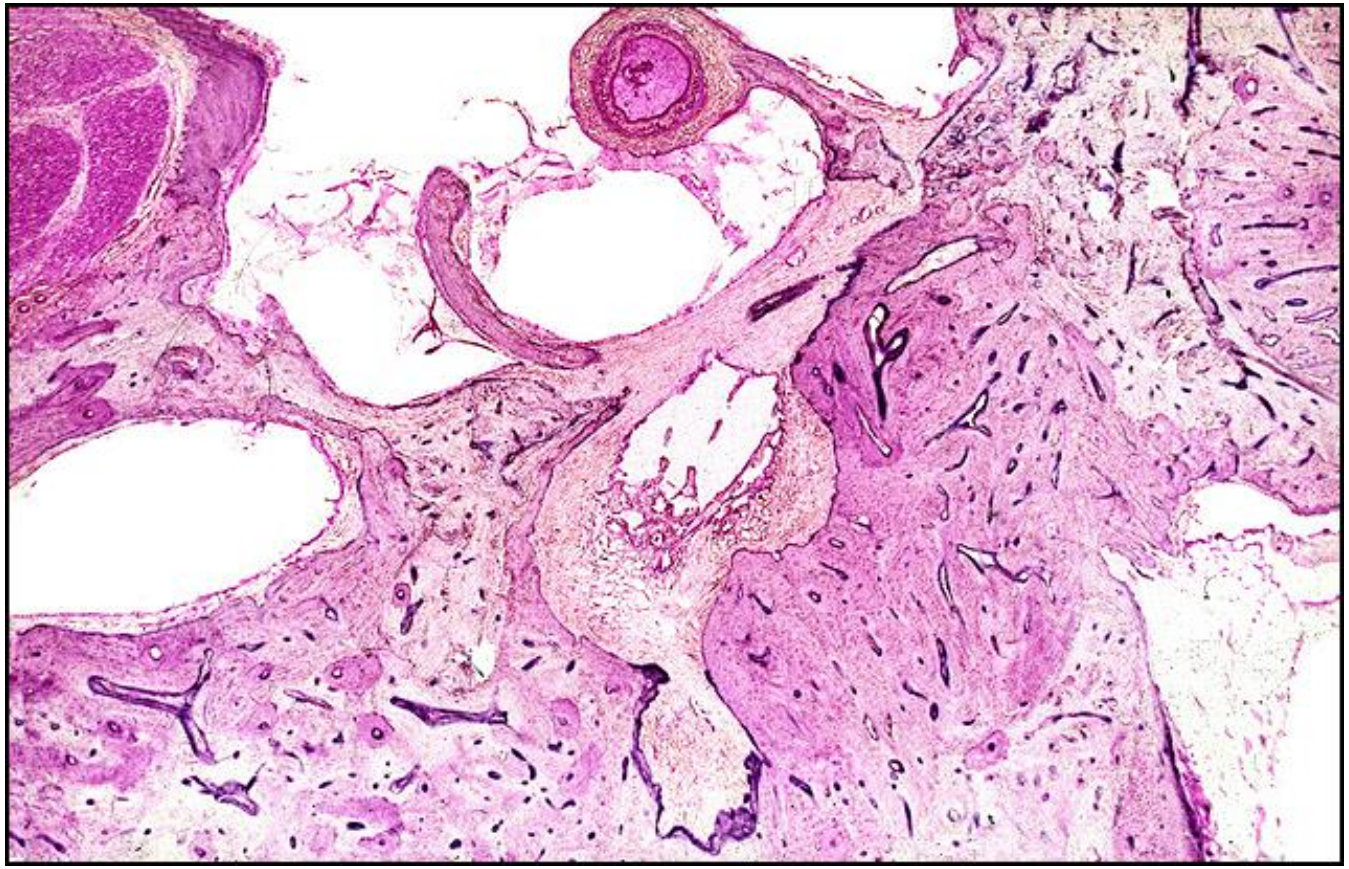
Chapter Section: Variform Anomalies External and Middle Ear / Persistent Stapedial Artery

TB Case Number: 325

Gender: Male

Otologic Diagnosis:

1. Multiple middle ear anomalies including:
    a) Obliteration of the middle ear with mesenchyme and bone
    b) Ossicular anomalies
    c) Congenital cholesteatoma medial to the tympanic membrane
    d) Partial development of the footplate
2. Normal bony and membranous labyrinth



Persistent Stapedial Artery, L-291

Title: Persistent Stapedial Artery, L-291

Chapter: Developmental Defects

Chapter Section: Persistent Stapedial Artery, Variform Anomalies External and Middle Ear

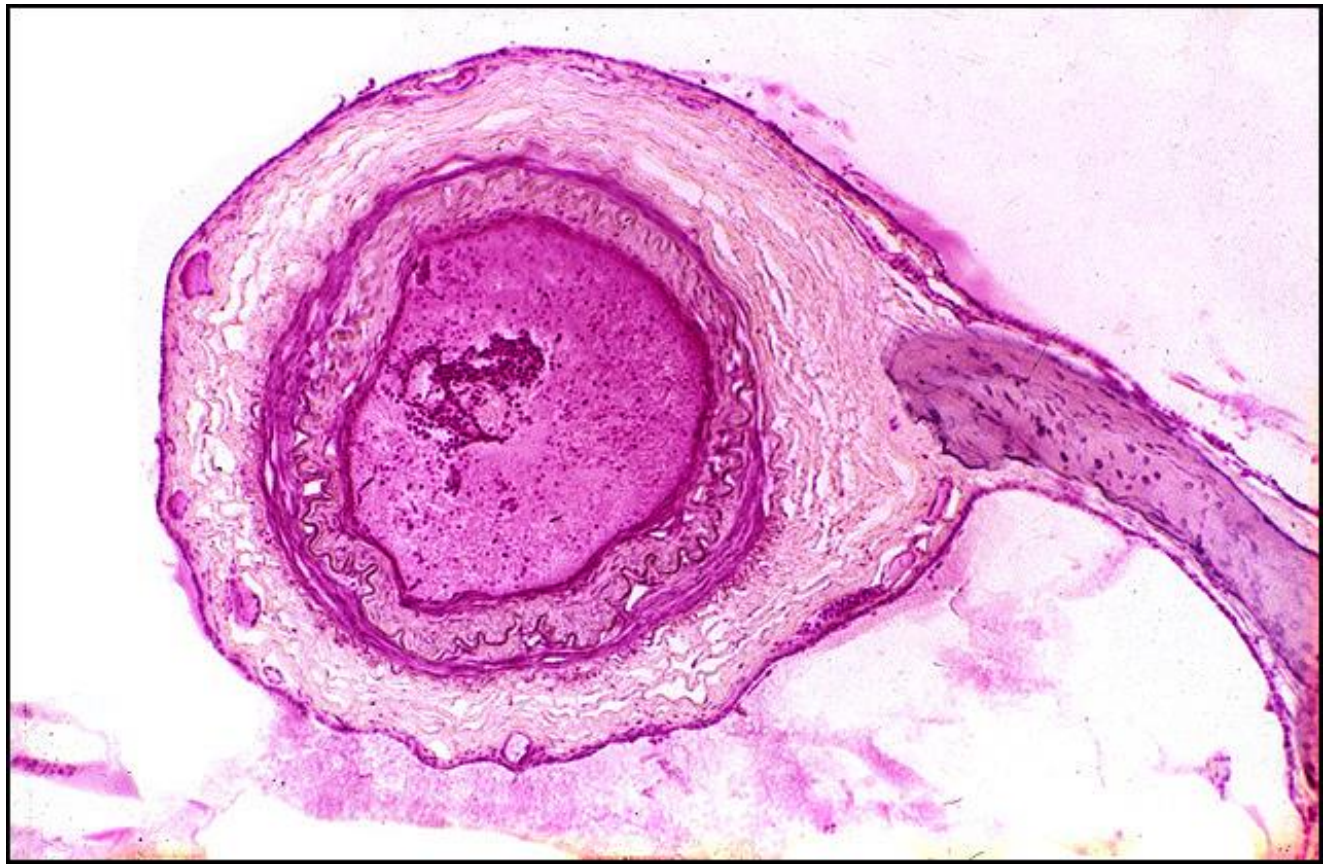
TB Case Number: 180

Gender: Male

Age (yrs.): 85

Otologic Diagnosis:

1. Labyrinthitis, remote, type undetermined, bilateral
2. Sensorineural hearing loss, total, bilateral
3. Labyrinthitis ossificans, severe, cochlear and vestibular, childhood labyrinthitis, presumptive, bilateral
4. Membranous labyrinth, atrophy, total



Persistent Stapedial Artery, L-291

Title: Persistent Stapedial Artery, L-291

Chapter: Developmental Defects

Chapter Section: Persistent Stapedial Artery, Variform Anomalies External and Middle Ear

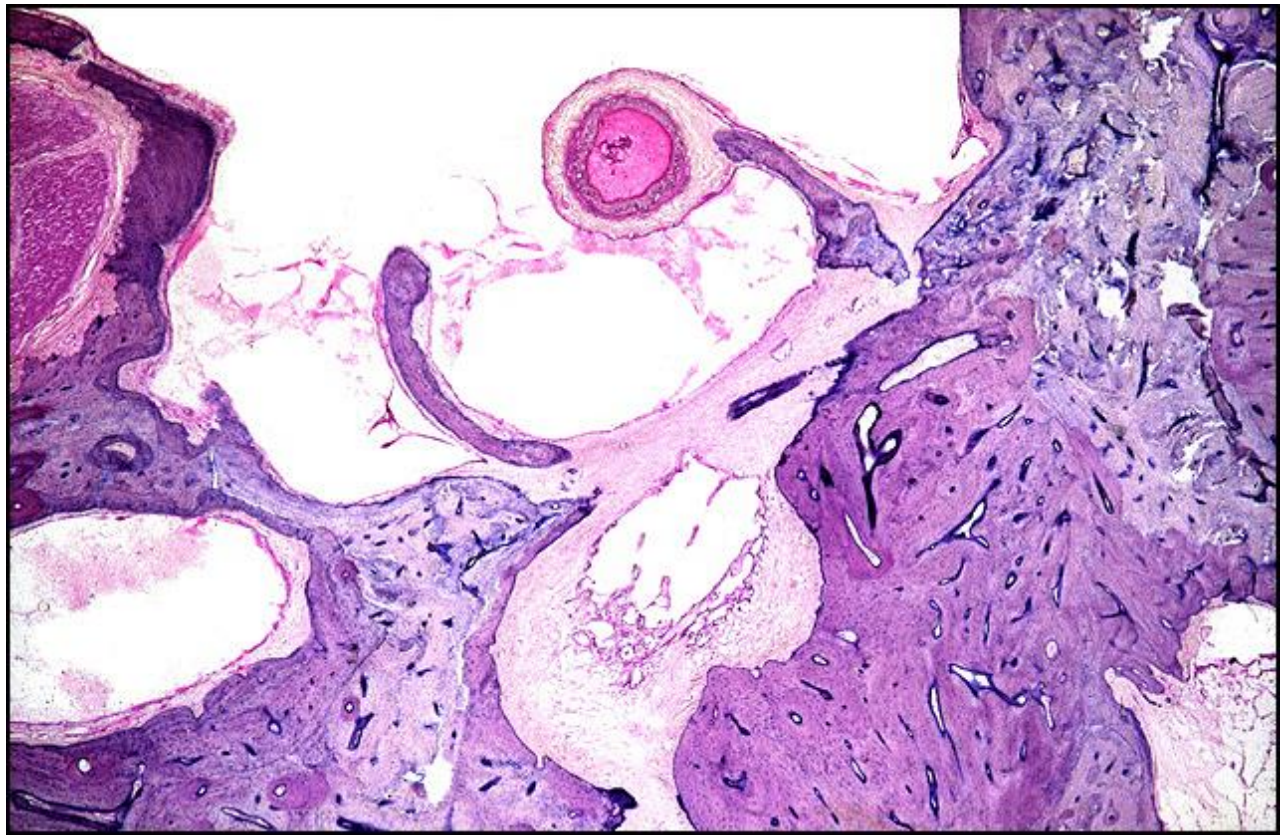
TB Case Number: 180

Gender: Male

Age (yrs.): 85

Otologic Diagnosis:

1. Labyrinthitis, remote, type undetermined, bilateral
2. Sensorineural hearing loss, total, bilateral
3. Labyrinthitis ossificans, severe, cochlear and vestibular, childhood labyrinthitis, presumptive, bilateral
4. Membranous labyrinth, atrophy, total



Persistent Stapedial Artery, L-382

Title: Persistent Stapedial Artery, L-382

Chapter: Developmental Defects

Chapter Section: Persistent Stapedial Artery, Variform Anomalies External and Middle Ear

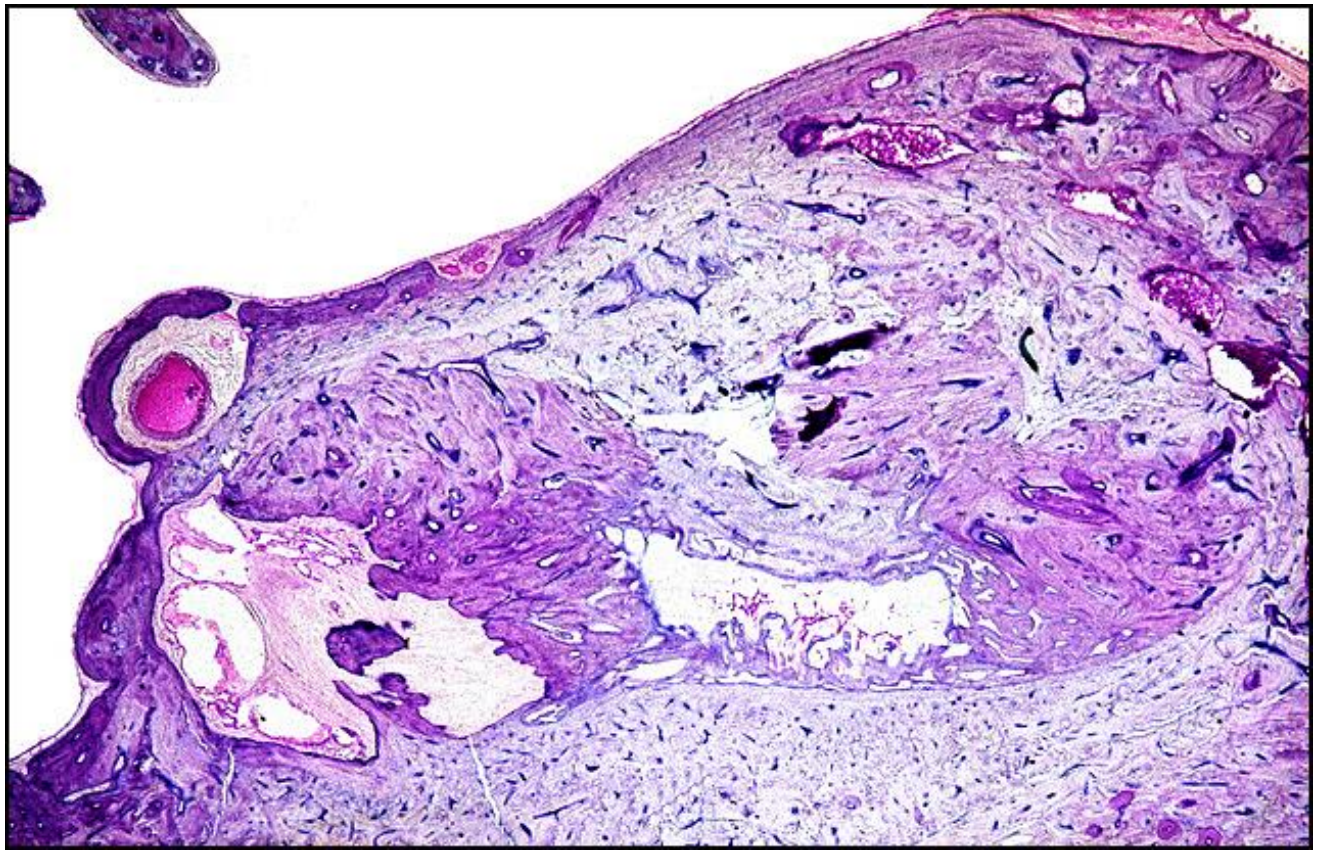
TB Case Number: 180

Gender: Male

Age (yrs.): 85

Otologic Diagnosis:

1. Labyrinthitis, remote, type undetermined, bilateral
2. Sensorineural hearing loss, total, bilateral
3. Labyrinthitis ossificans, severe, cochlear and vestibular, childhood labyrinthitis, presumptive, bilateral
4. Membranous labyrinth, atrophy, total



Schwannosis, R-101.2

Title: Schwannosis, R-101.2

Chapter: Developmental Defects

Chapter Section: Ectopic Muscle, Schwannosis, Variform Anomalies External and Middle Ear

TB Case Number: 1014

Comments: Nodular swelling of facial nerve in IAC. Benign hyperplasia of Schwann cells. Consistent with schwannosis. 98-year-old man with longstanding Meniere's syndrome for 50 years.

Gender: Male

Age (yrs.): 99

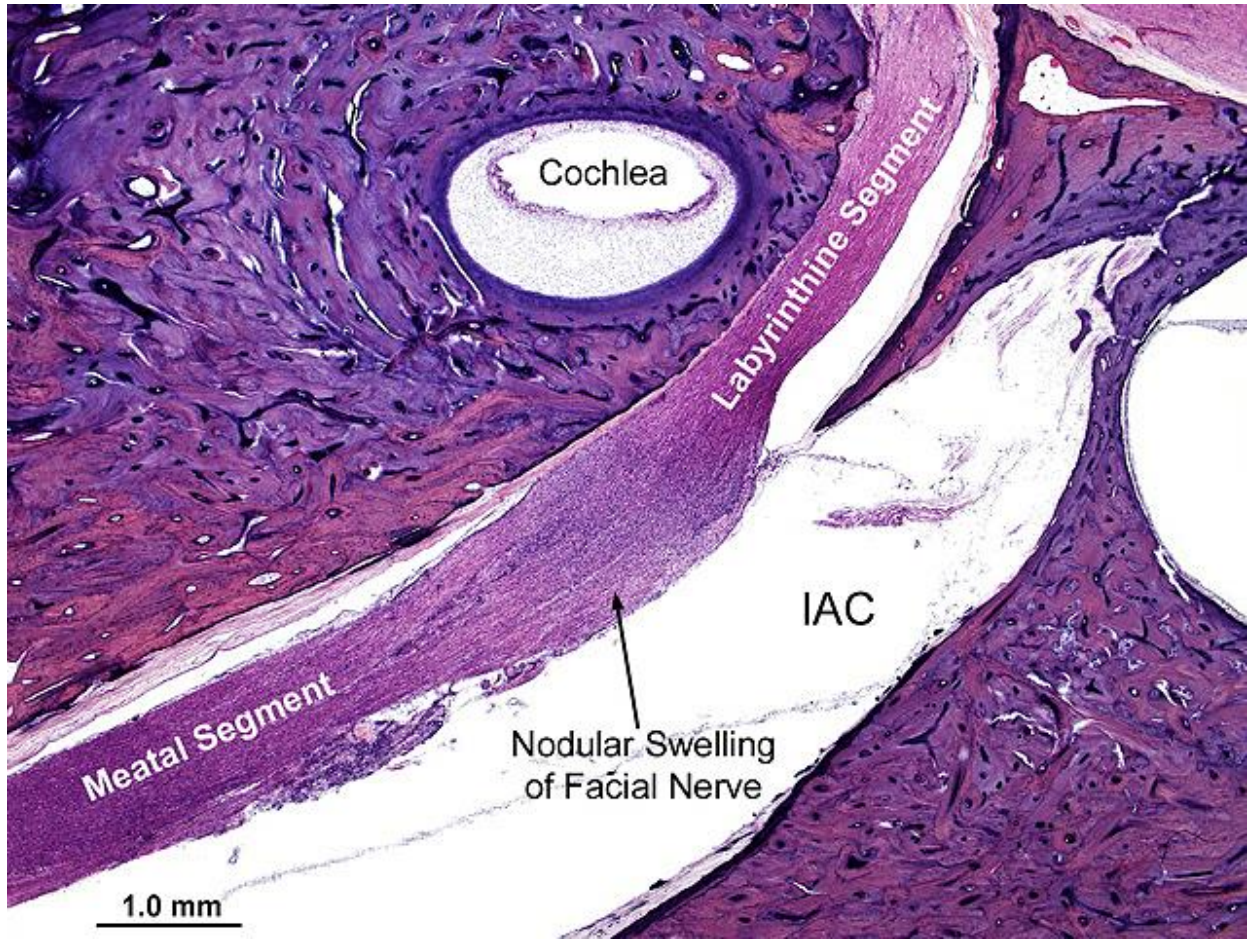
Otologic Diagnosis:

Clinical:

1. Meniere’s syndrome, left ear
2. Profound sensorineural hearing loss, bilateral

Histopathologic Diagnoses:

Endolymphatic hydrops, severe and diffuse, involving cochlea, saccule, utricle, and all three ampullae, bilateral



Schwannosis, R-101.1

Title: Schwannosis, R-101.1

Chapter: Developmental Defects

Chapter Section: Ectopic Muscle, Schwannosis, Variform Anomalies External and Middle Ear

TB Case Number: 1014

Comments: Nodular swelling of facial nerve in IAC. Benign hyperplasia of Schwann cells. Consistent with schwannosis. 98-year-old man with longstanding Meniere's syndrome for 50 years.

Gender: Male

Age (yrs.): 99

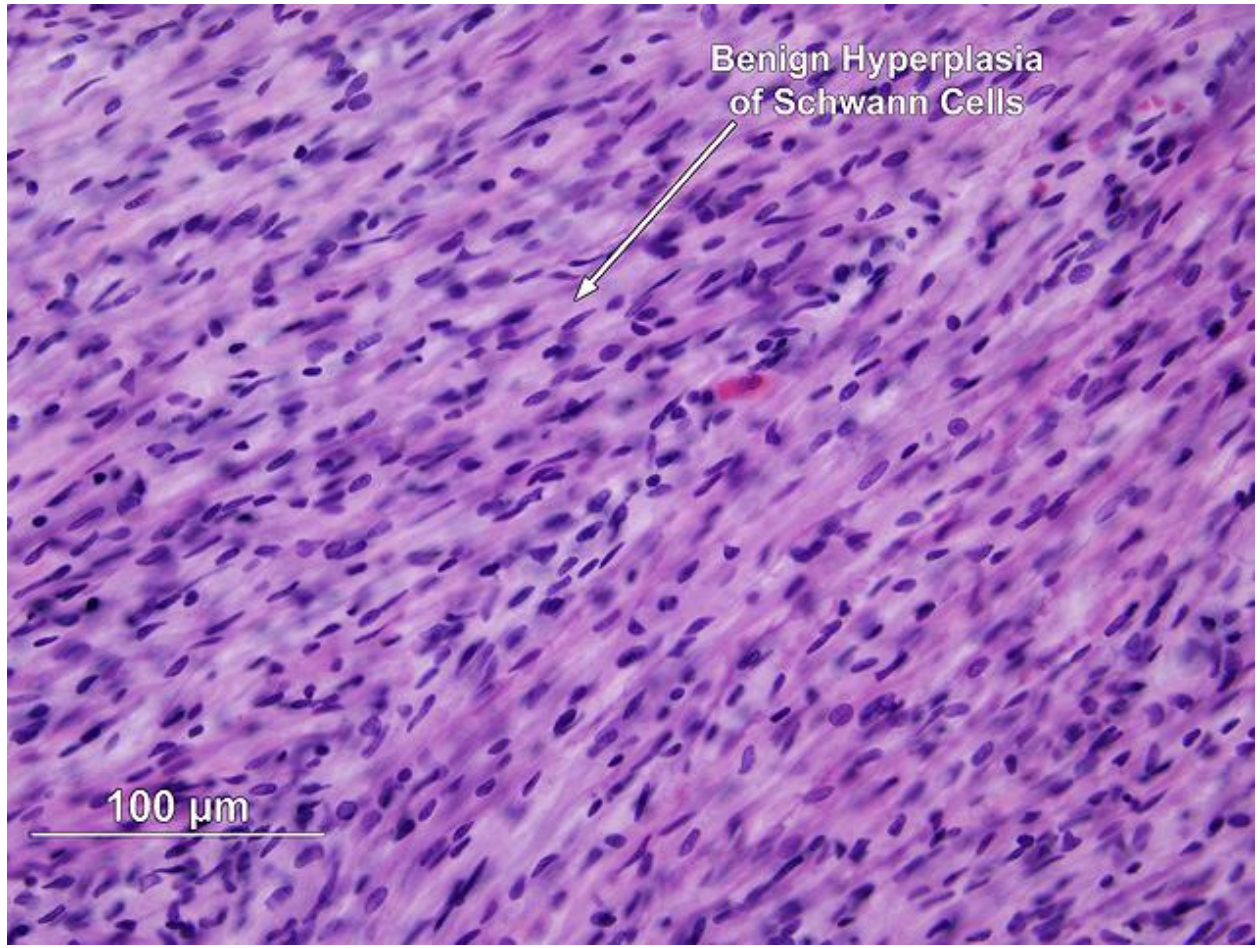
Otologic Diagnosis:

Clinical:

1. Meniere’s syndrome, left ear
2. Profound sensorineural hearing loss, bilateral

Histopathologic Diagnoses:

Endolymphatic hydrops, severe and diffuse, involving cochlea, saccule, utricle, and all three ampullae, bilateral



Schwannosis, L-81

Title: Schwannosis, L-81

Chapter: Developmental Defects

Chapter Section: Ectopic Muscle, Schwannosis, Variform Anomalies External and Middle Ear

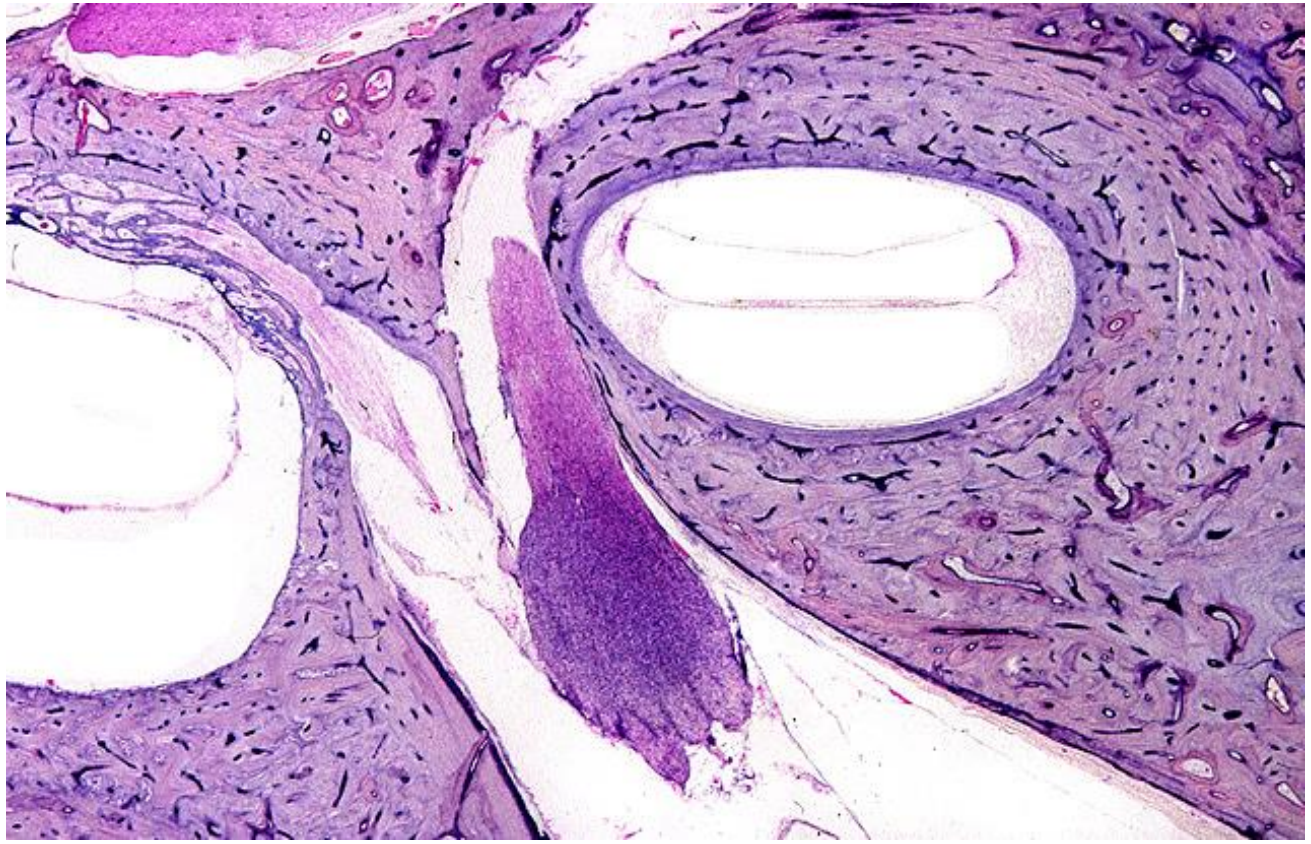
TB Case Number: 197

Gender: Male

Age (yrs.): 77

Otologic Diagnosis:

1. Labyrinthitis, acute, putative, viral, limited to cochlea, remote, right
2. Organ of Corti, degeneration, secondary to cochlear labyrinthitis, right
3. Tectorial membrane, atrophy, severe, secondary to cochlear labyrinthitis, right
4. Retrograde neuron



Schwannosis, R-71

Title: Schwannosis, R-71

Chapter: Developmental Defects

Chapter Section: Ectopic Muscle, Schwannosis, Variform Anomalies, External and Middle Ear

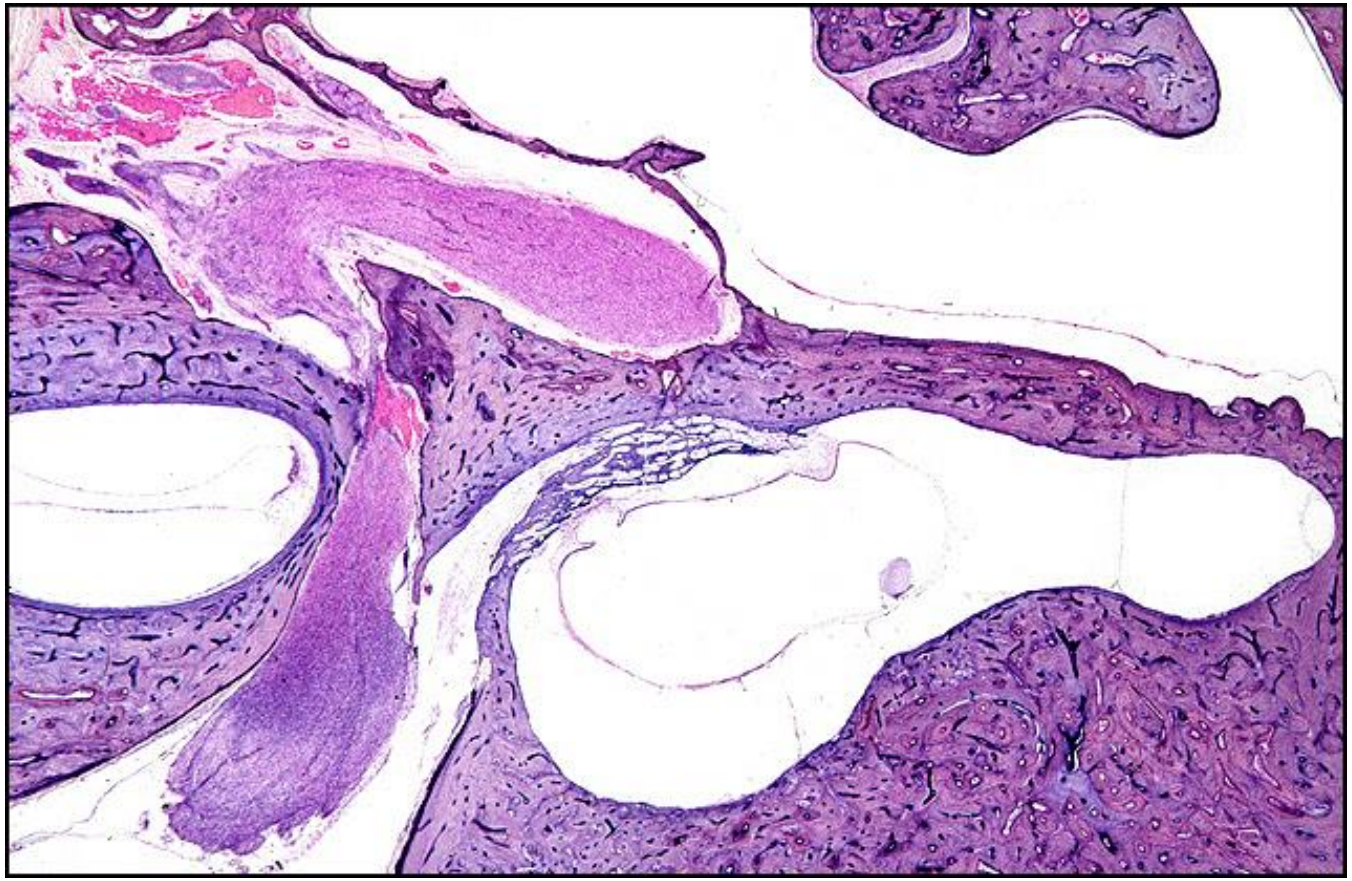
TB Case Number: 197

Gender: Male

Age (yrs.): 77

Otologic Diagnosis:

1. Labyrinthitis, acute, putative, viral, limited to cochlea, remote, right
2. Organ of Corti, degeneration, secondary to cochlear labyrinthitis, right
3. Tectorial membrane, atrophy, severe, secondary to cochlear labyrinthitis, right
4. Retrograde neuron



Bifurcation Facial Nerve, L-11

Title: Bifurcation Facial Nerve, L-11

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies, External and Middle Ear

TB Case Number: 641

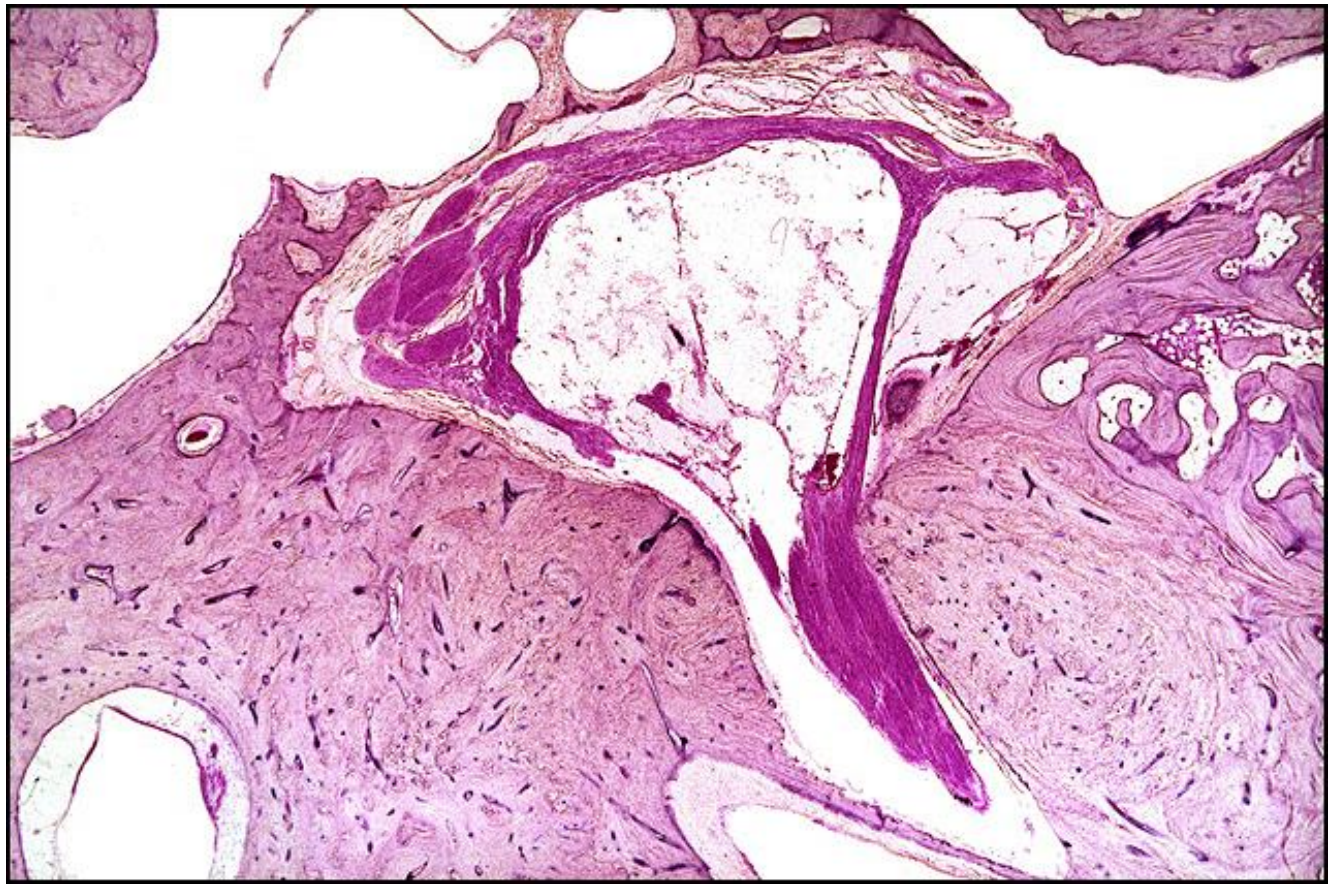
Gender: Female

Age (yrs.): 45

Otologic Diagnosis:

Congenital anomalies consisting of:

1. Osseous spiral lamina, defective, right
2. Posterior semicircular canal, missing, left
3. Crista, horizontal canal, aplastic, left
4. Cochlear nerve, degeneration, more severe on right, bilateral



Bifurcation Facial Nerve, L-56

Title: Bifurcation Facial Nerve, L-56

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies, External and Middle Ear

TB Case Number: 641

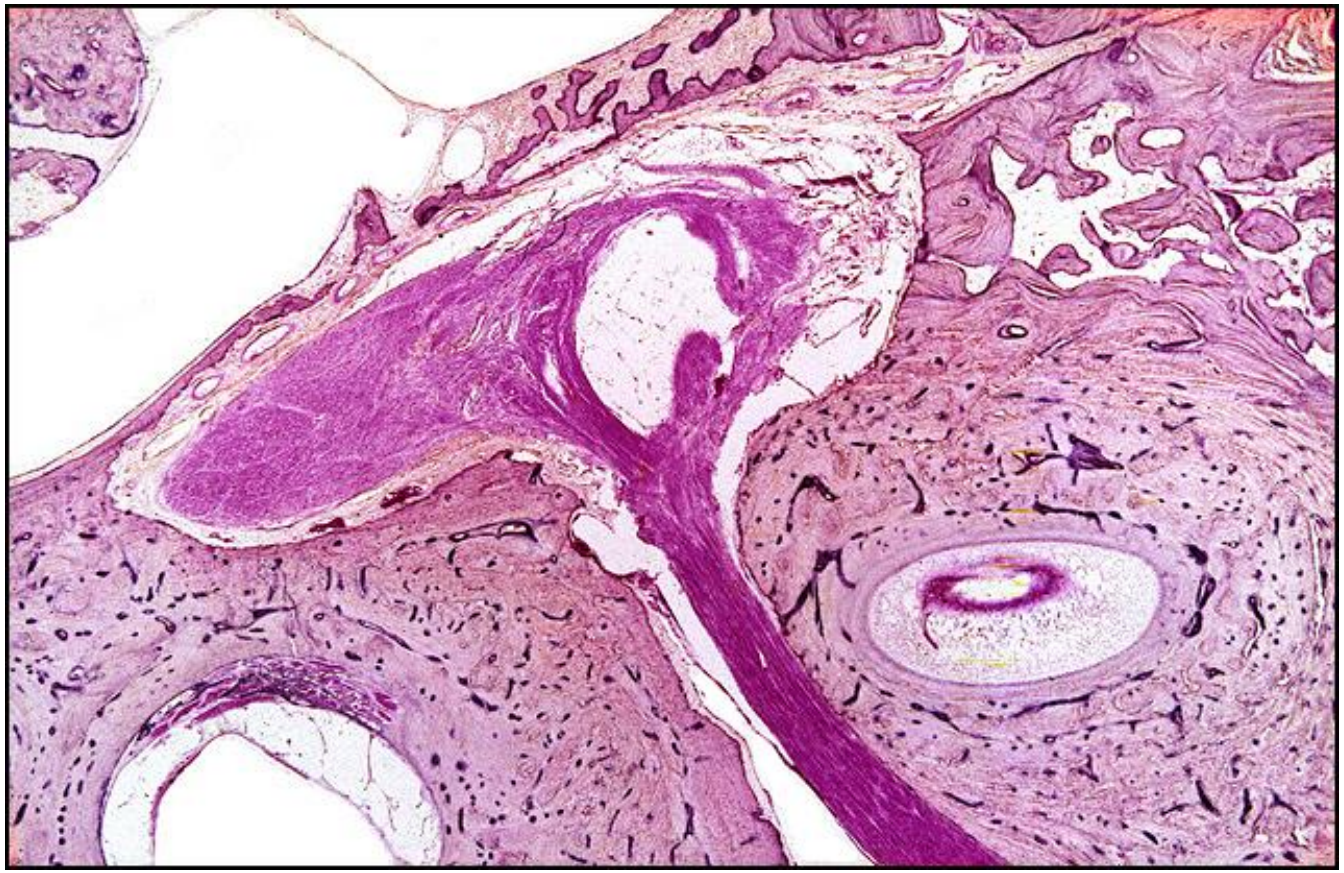
Gender: Female

Age (yrs.): 45

Otologic Diagnosis:

Congenital anomalies consisting of:

1. Osseous spiral lamina, defective, right
2. Posterior semicircular canal, missing, left
3. Crista, horizontal canal, aplastic, left
4. Cochlear nerve, degeneration, more severe on right, bilateral



Facial Hiatus, L-60

Title: Facial Hiatus, L-60

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies, External and Middle Ear

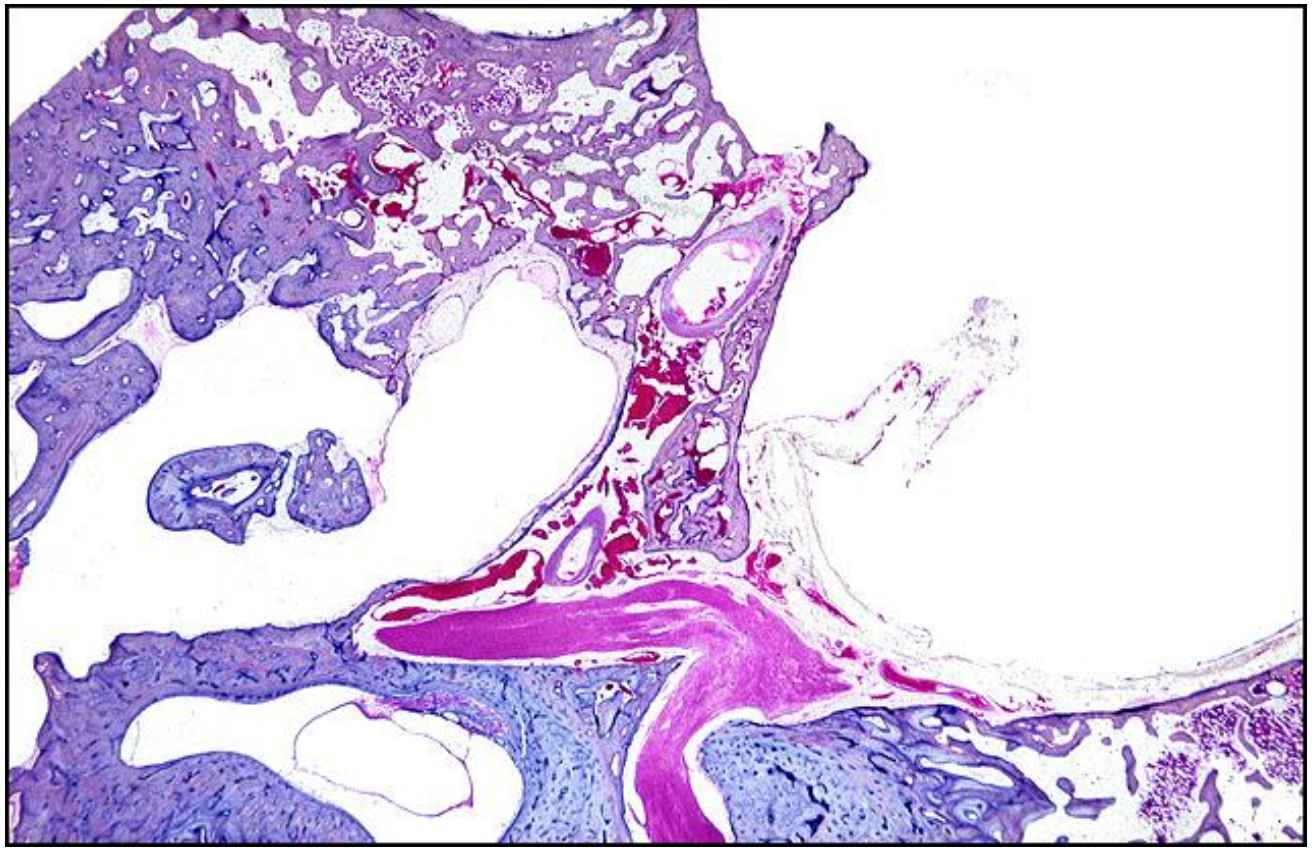
TB Case Number: 549

Gender: Female

Age (yrs.): 76

Otologic Diagnosis:

1. Normal middle ear and inner ear structures (no explanation for the patient's vestibular symptoms)
2. Persistent stapedial artery, left ear
3. Mucosal invagination into canal of the inferior tympanic nerve, left ear



Geniculate Ganglion, L-31

Title: Geniculate Ganglion, L-31

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies External and Middle Ear

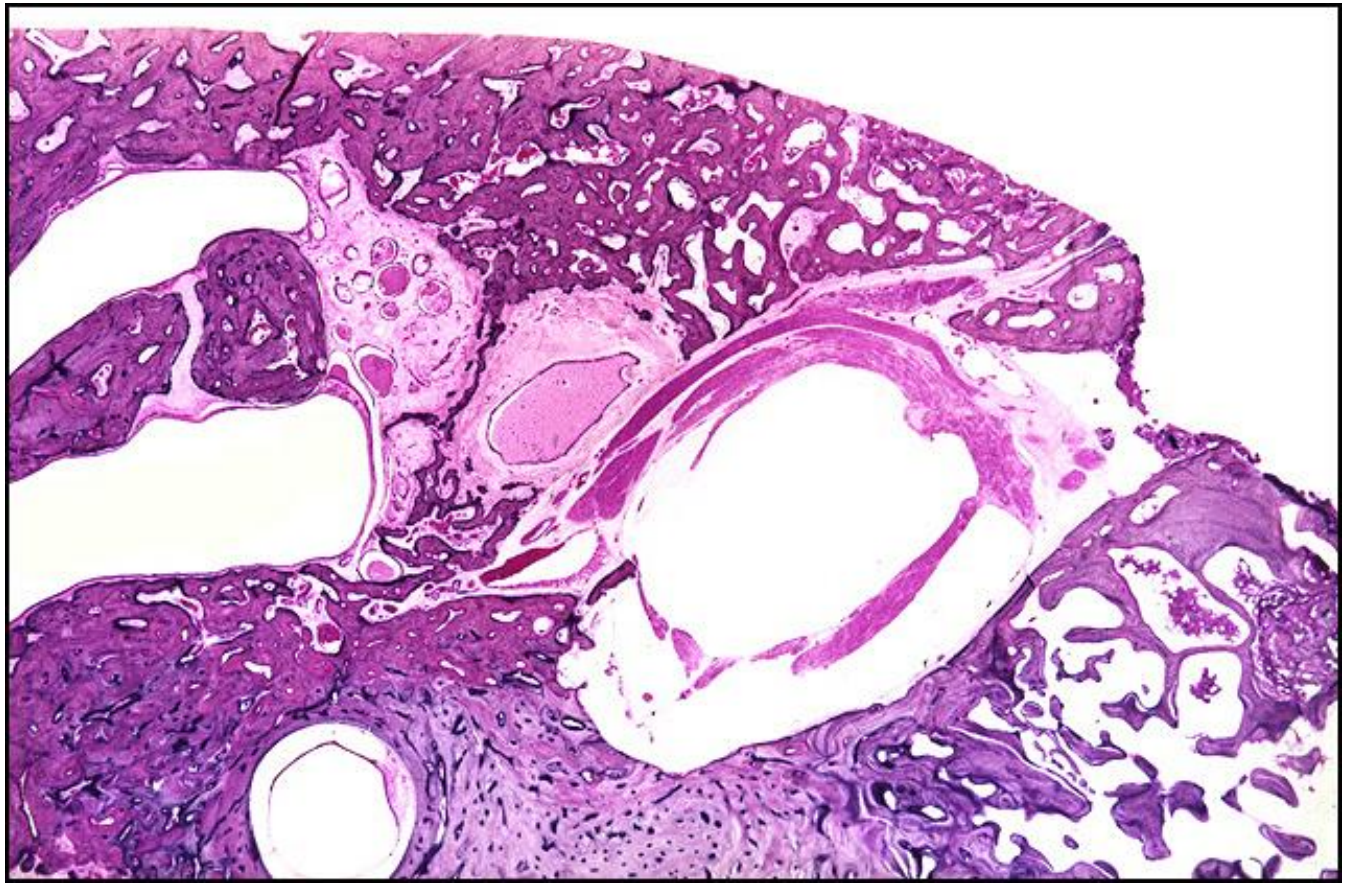
TB Case Number: 839

Gender: Female

Age (yrs.): 75

Otologic Diagnosis:

1. Otitis media, chronic, showing destruction of tympanic membrane, ossicles, and epidermalization of mesotympanum, left
2. Fibrocystic and osseous sclerosis, mastoid, left
3. Spiral ligament, atrophy, severe, left
4. Anatomic variant



Facial Nerve, L-21

Title: Facial Nerve, L-21

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies, External and Middle Ear

TB Case Number: 998

Comments: CSF space extension to geniculate ganglion

Gender: Female

Age (yrs.): 91

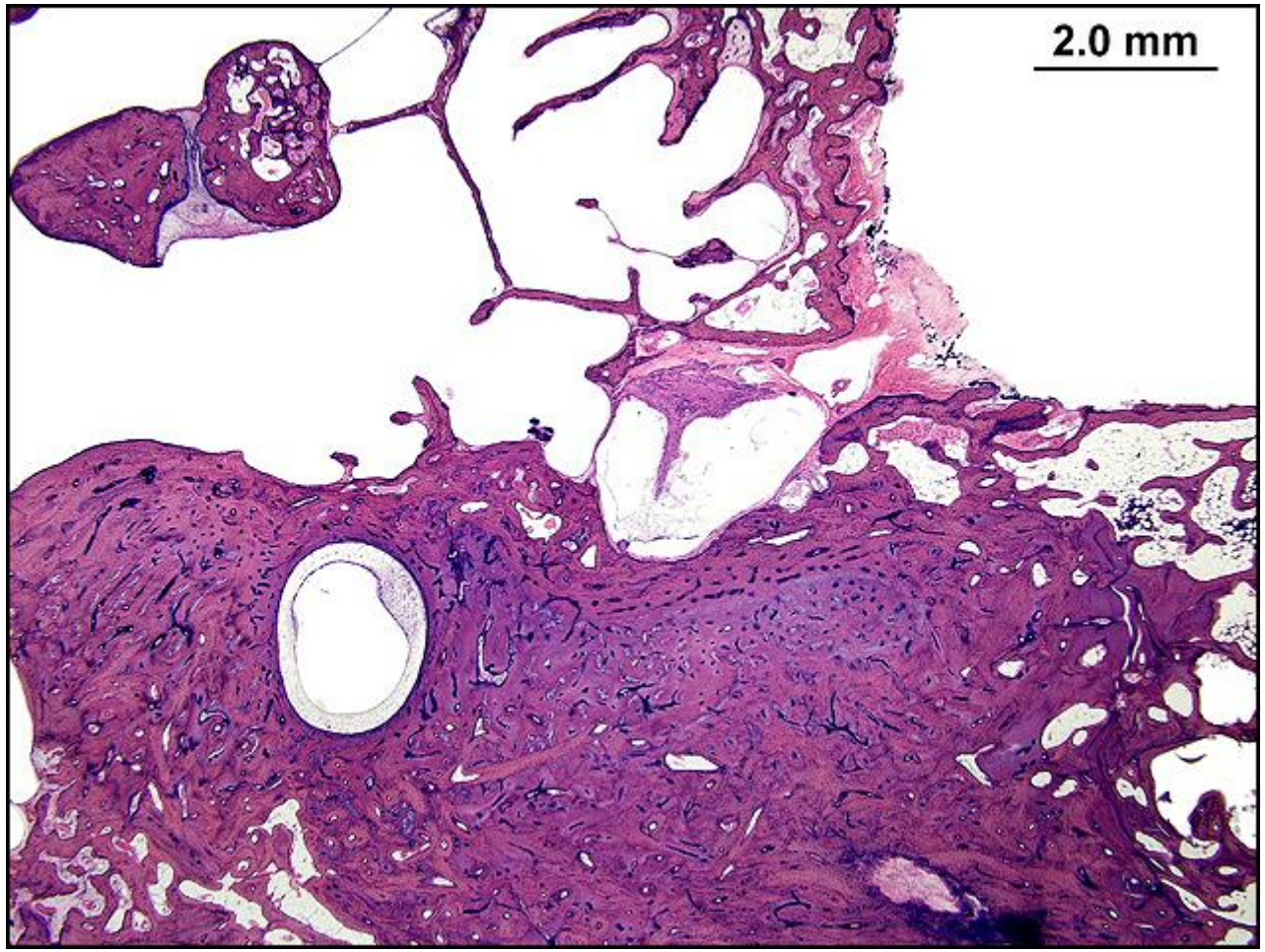
Otologic Diagnosis:

Clinical:

Progressive hearing loss, bilateral, presumed sensorineural and of genetic etiology

Histological:

Mild cochlear degeneration (outer hair cell loss in basal turn, mild patchy strial atrophy, mild basal spiral ganglion cell loss, spiral ligament atrophy



Facial Nerve, L-21

Title: Facial Nerve, L-21

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies, External and Middle Ear

TB Case Number: 998

Comments: CSF space extension to geniculate ganglion

Gender: Female

Age (yrs.): 91

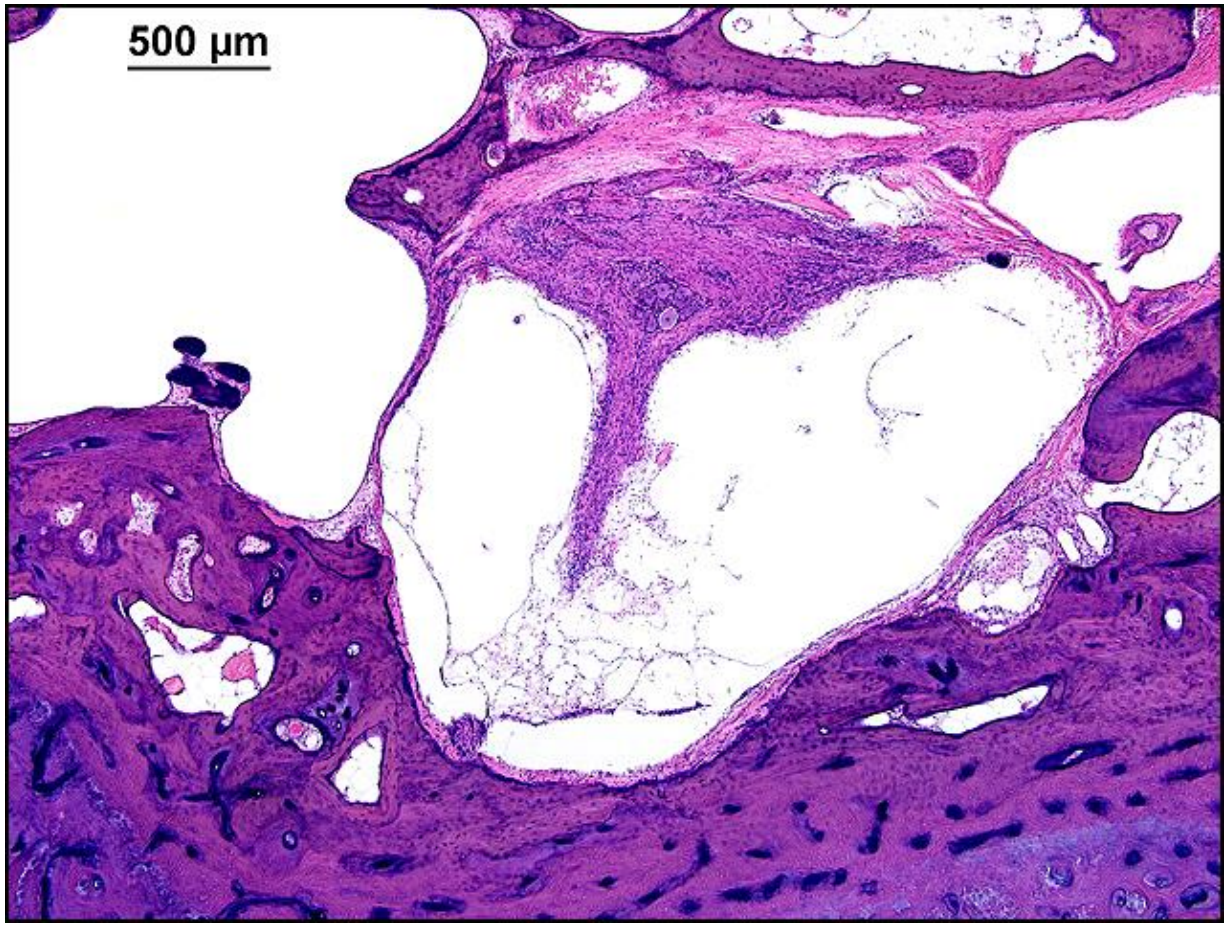
Otologic Diagnosis:

Clinical:

Progressive hearing loss, bilateral, presumed sensorineural and of genetic etiology

Histological:

Mild cochlear degeneration (outer hair cell loss in basal turn, mild patchy strial atrophy, mild basal spiral ganglion cell loss, spiral ligament atrophy



Facial Nerve, L-51

Title: Facial Nerve, L-51

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies, External and Middle Ear

TB Case Number: 998

Comments: CSF space extension to geniculate ganglion

Gender: Female

Age (yrs.): 91

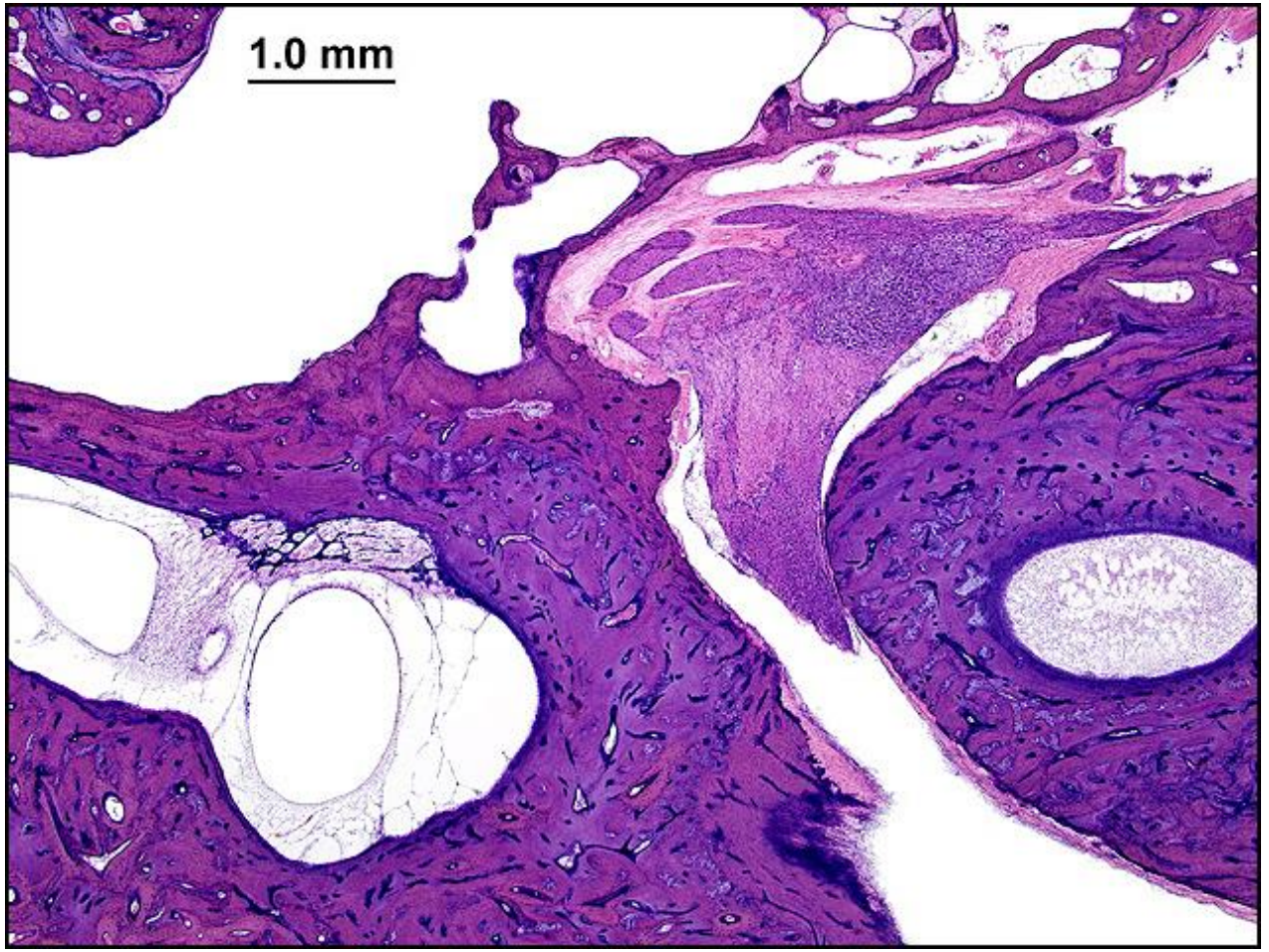
Otologic Diagnosis:

Clinical:

Progressive hearing loss, bilateral, presumed sensorineural and of genetic etiology

Histological:

Mild cochlear degeneration (outer hair cell loss in basal turn, mild patchy strial atrophy, mild basal spiral ganglion cell loss, spiral ligament atrophy



Large Facial Hiatus, L-41

Title: Large Facial Hiatus, L-41

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies, External and Middle Ear

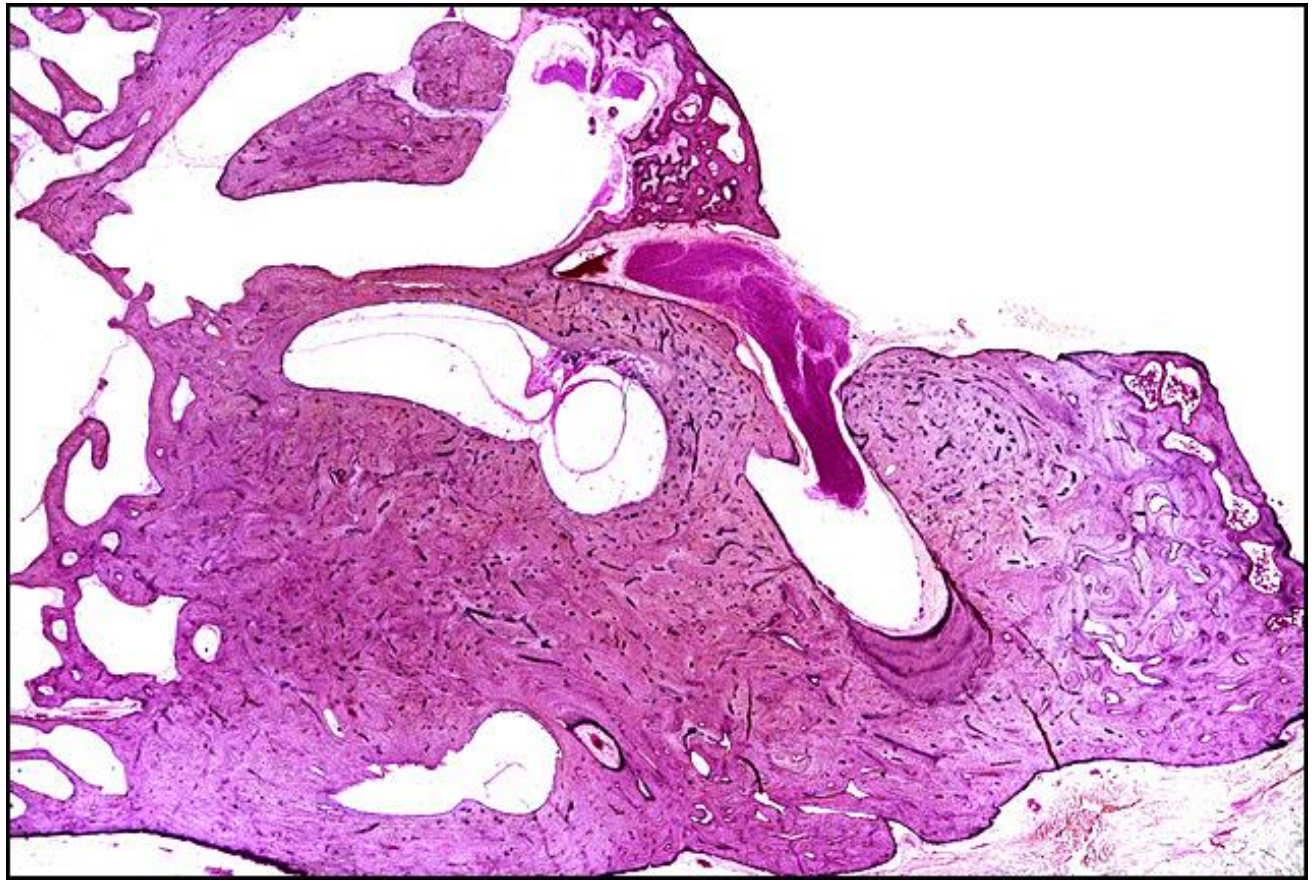
TB Case Number: 862

Gender: Female

Age (yrs.): 70

Otologic Diagnosis:

Neural presbycusis



Vein in Facial Canal, L-371

Title: Vein in Facial Canal, L-371

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies, External and Middle Ear

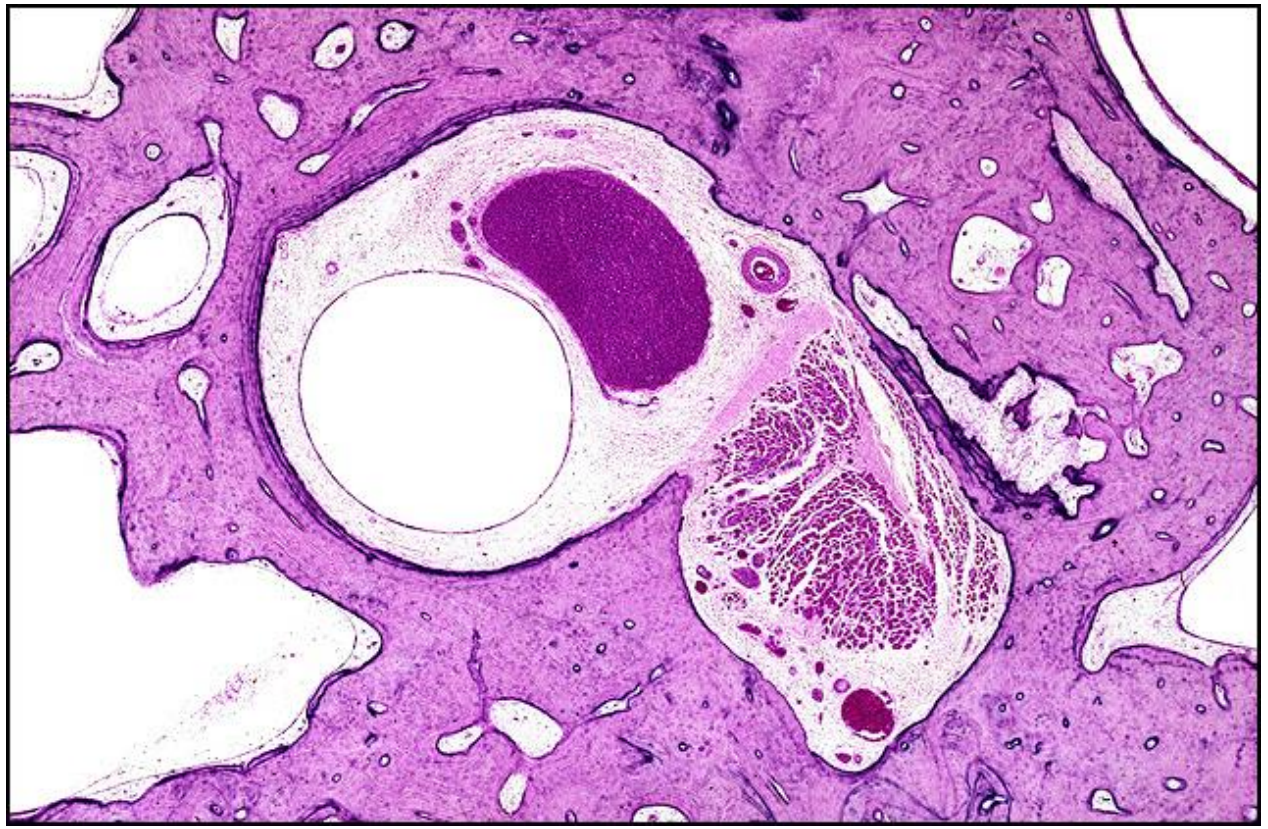
TB Case Number: 871

Gender: Male

Age (yrs.): 41

Otologic Diagnosis:

1. Normal temporal bone, right, excellent preservation
2. Normal temporal bone, left, compression artifact



Vein in Facial Canal, R-500

Title: Vein in Facial Canal, R-500

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies External and Middle Ear

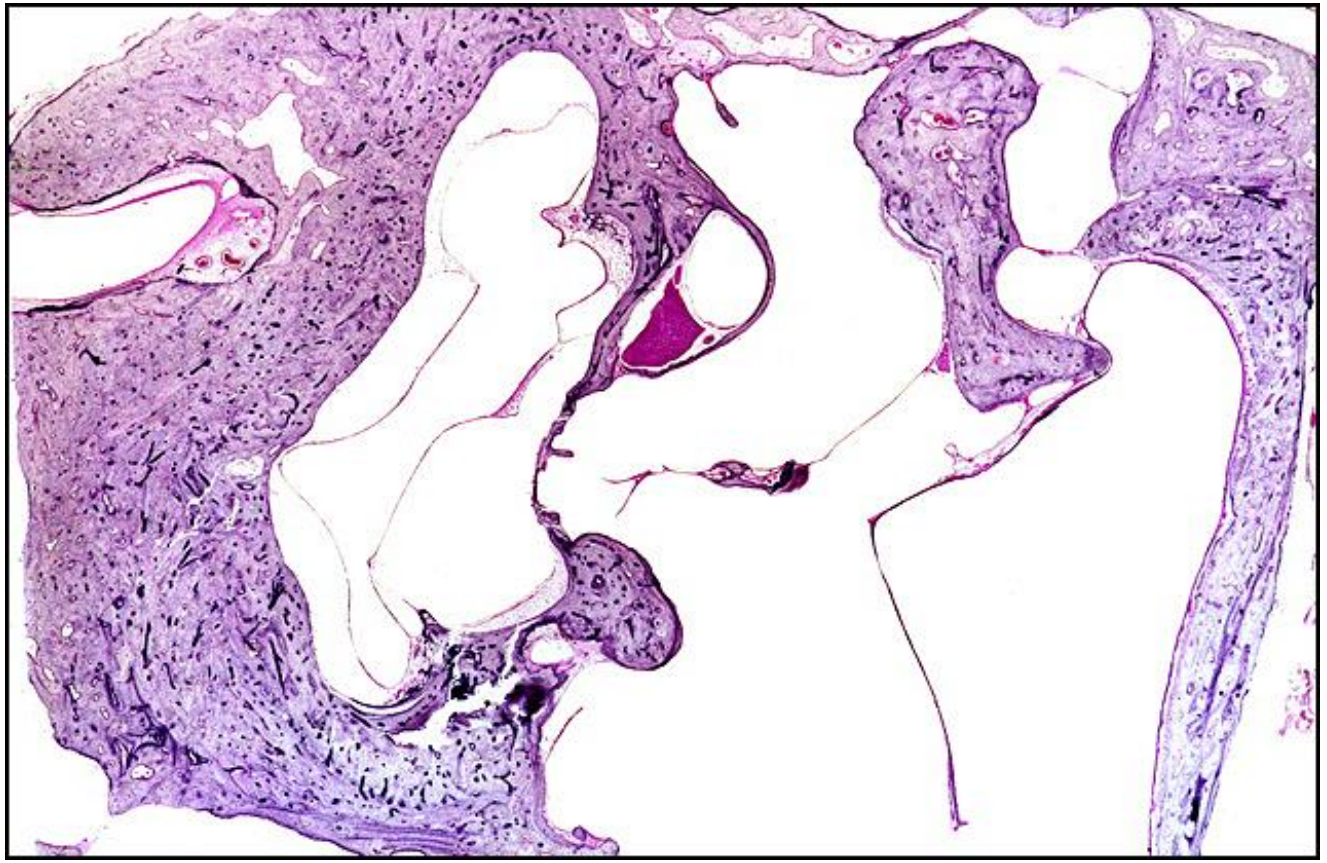
TB Case Number: 465

Gender: Female

Age (yrs.): 87

Otologic Diagnosis:

1. Stria vascularis, atrophy, severe, bilateral
2. Tympanic membrane, perforation, bilateral
3. Malleus, ankylosis to tegmen tympani, right
4. Fallopian canal, large vein present, presumably in continuity with dural vein, bilateral



Facial Nerve Anomaly, R-80

Title: Facial Nerve Anomaly, R-80

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies External and Middle Ear

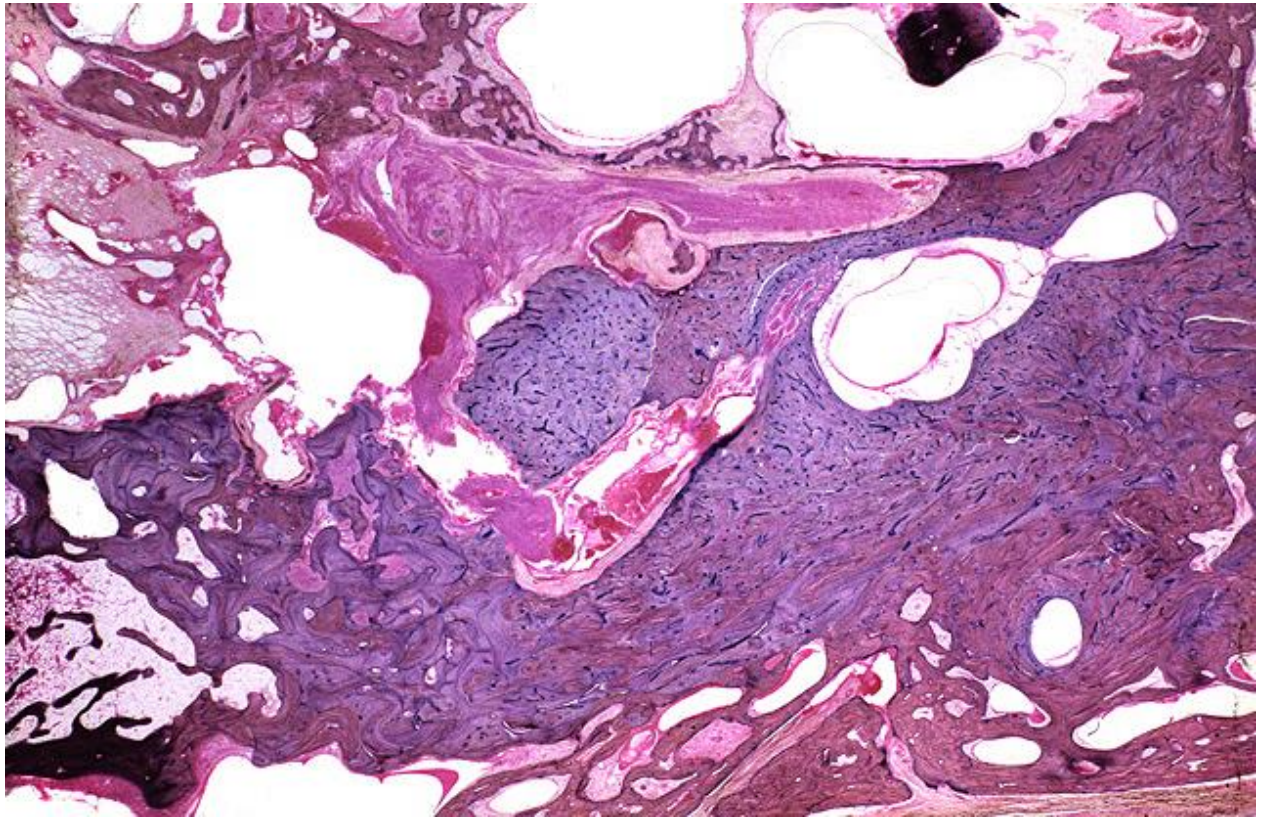
TB Case Number: 221

Gender: Male

Age (yrs.): 56

Otologic Diagnosis:

1. Schwannoma, cerebellopontine angle, clinical, left
2. Hemotympanum, postoperative, schwannoma, left
3. Subarachnoid hemorrhage, postoperative, bilateral
4. Membranous labyrinth, degeneration, schwannoma, left
5. Organ of Corti, degeneration, schwannoma



Facial Nerve Dehiscence at Oval Window, R-166

Title: Facial Nerve Dehiscence at Oval Window, R-166

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve, Variform Anomalies External and Middle Ear

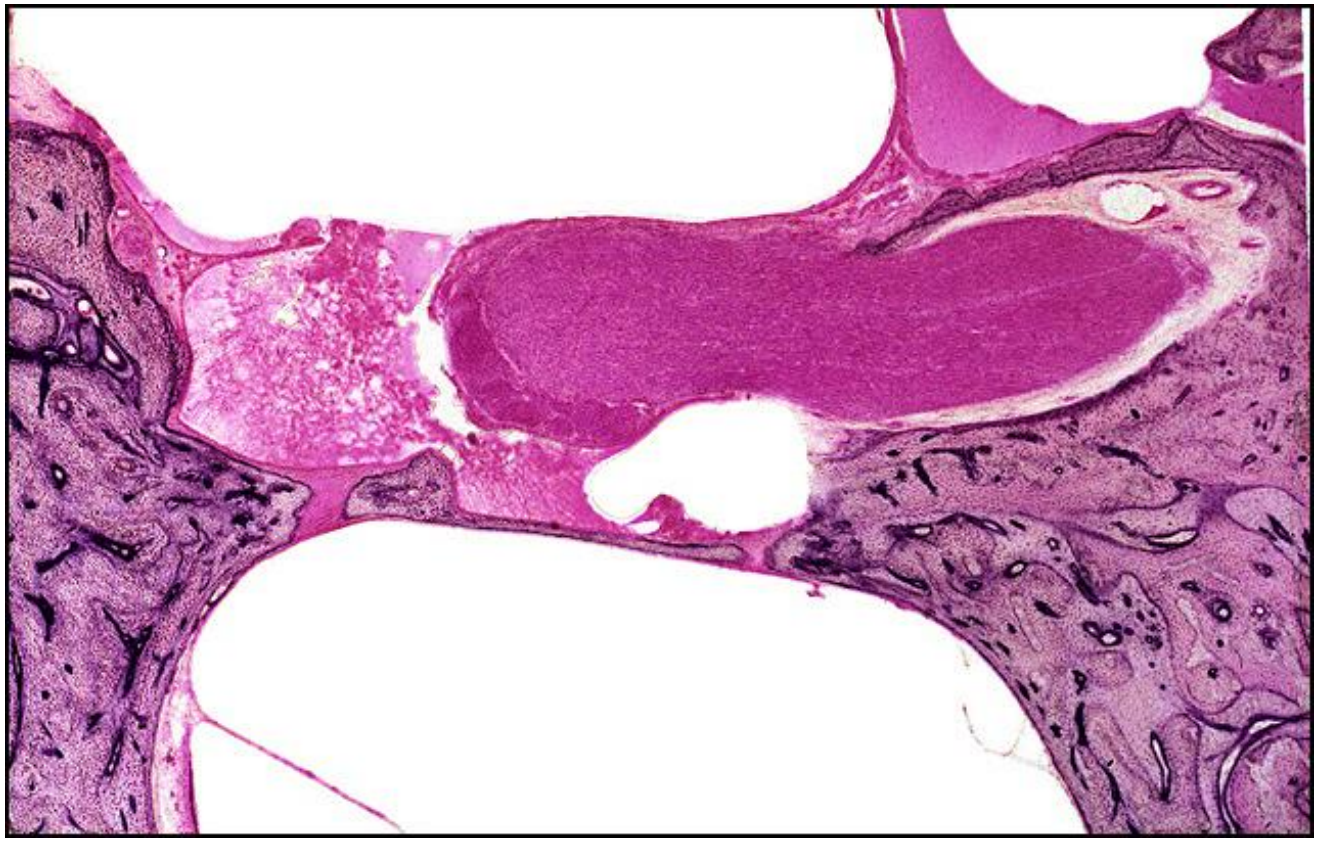
TB Case Number: 310

Gender: Male

Age (yrs.): 51

Otologic Diagnosis:

1. Labyrinthitis ossificans, perilymphatic space, posterior canal, right
2. Semicircular canal, posterior, osseous, obliteration, right
3. Facial nerve, dehiscent, oval window area, right
4. Membranous labyrinth, normal, bilateral
5. Artifact, preparation



Facial Nerve Anomaly, R-371

Title: Facial Nerve Anomaly, R-371

Chapter: Developmental Defects

Chapter Section: Anatomic Variants, Facial Nerve

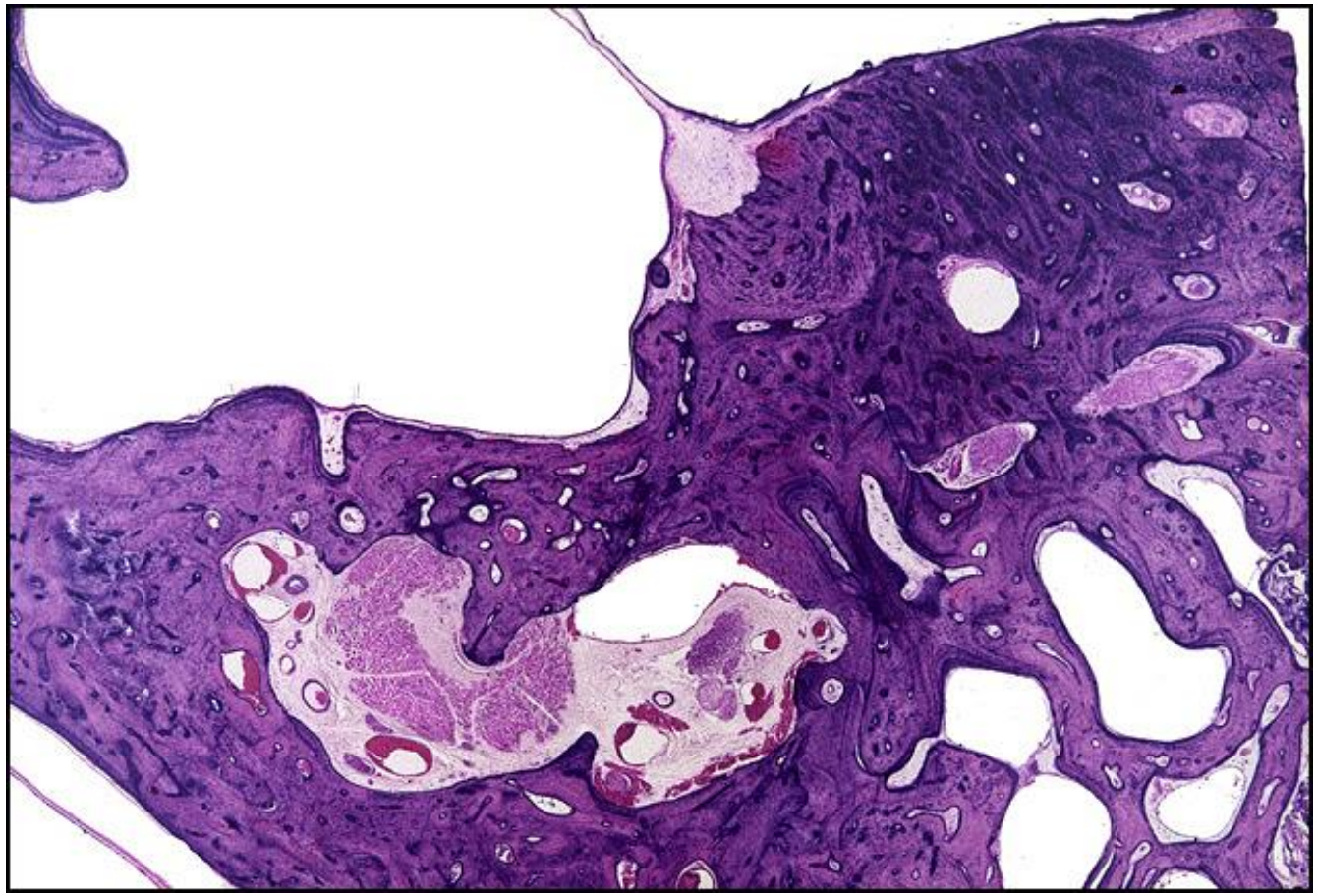
TB Case Number: 401

Gender: Female

Age (yrs.): 61

Otologic Diagnosis:

1. Otosclerosis, bilateral
2. Stapedectomy with fat-wire prosthesis, right
3. Degeneration of chorda tympani nerve
4. Anomaly facial nerve, right



Arachnoid Granulation, L-311.1

Title: Arachnoid Granulation, L-311.1

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

TB Case Number: 1024

Comments: 94 yr old female with occult arachnoid granulation against tegmen tympani (middle cranial fossa)

Gender: Female

Age (yrs.): 95

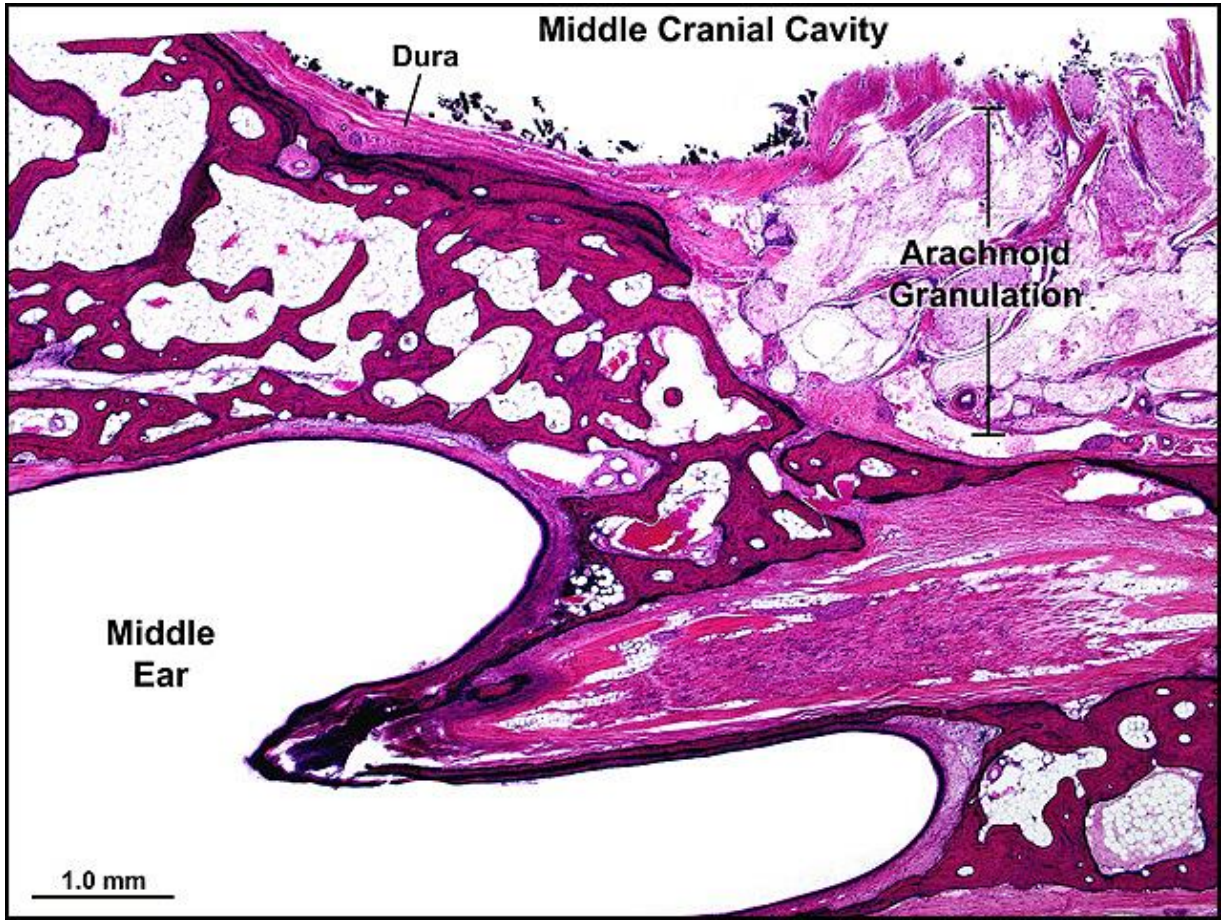
Otologic Diagnosis:

Clinical:

Adult onset, progressive, bilateral sensorineural hearing loss, probably genetic in etiology

Histopathologic:

Stria vascularis atrophy, near total, affecting entire cochlear duct, bilateral, probably genetically determined



Arachnoid Granulation, L-311.2

Title: Arachnoid Granulation, L-311.2

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

TB Case Number: 1024

Comments: 94 yr. old female with occult arachnoid granulation against tegmen tympani (middle cranial fossa)

Gender: Female

Age (yrs.): 95

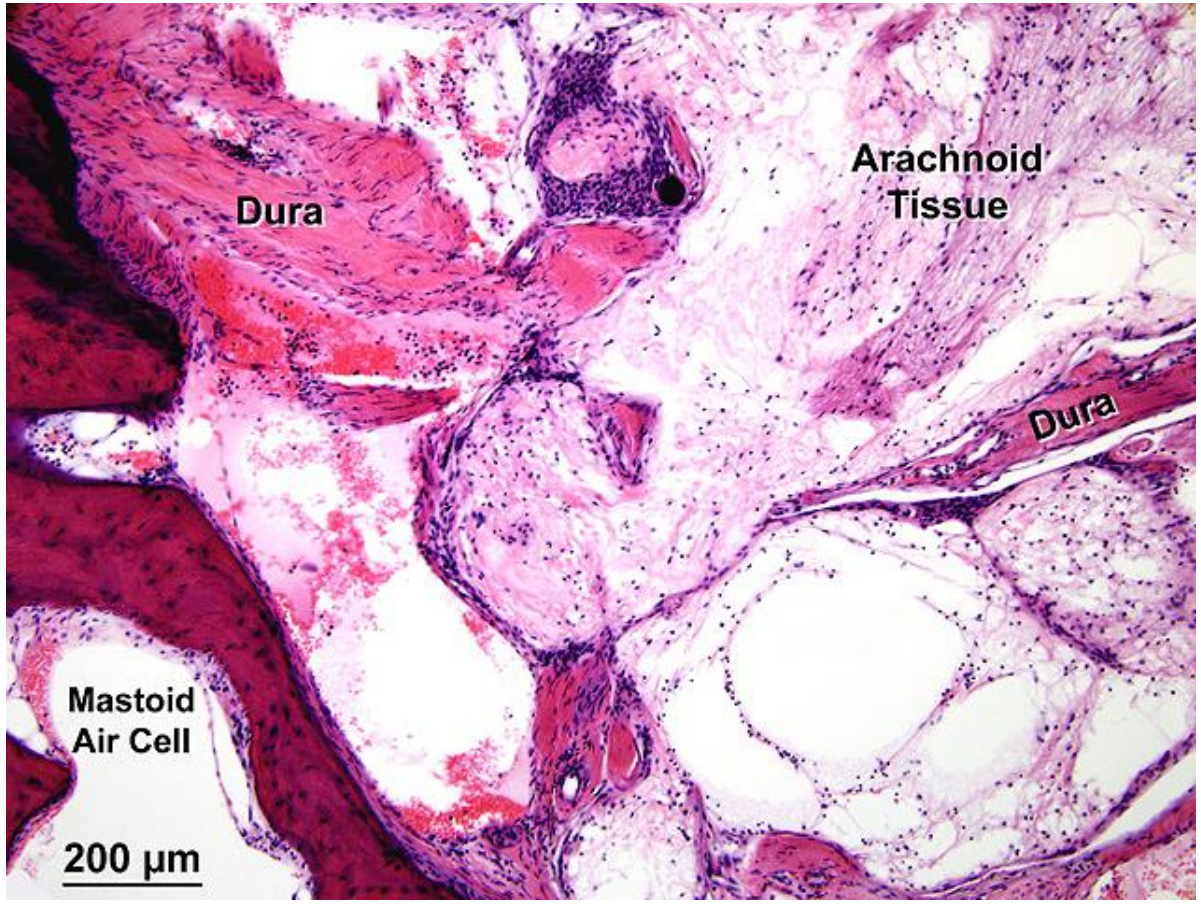
Otologic Diagnosis:

Clinical:

Adult onset, progressive, bilateral sensorineural hearing loss, probably genetic in etiology

Histopathologic:

Stria vascularis atrophy, near total, affecting entire cochlear duct, bilateral, probably genetically determined



Arachnoid Granulation, L-100

Title: Arachnoid Granulation, L-100

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

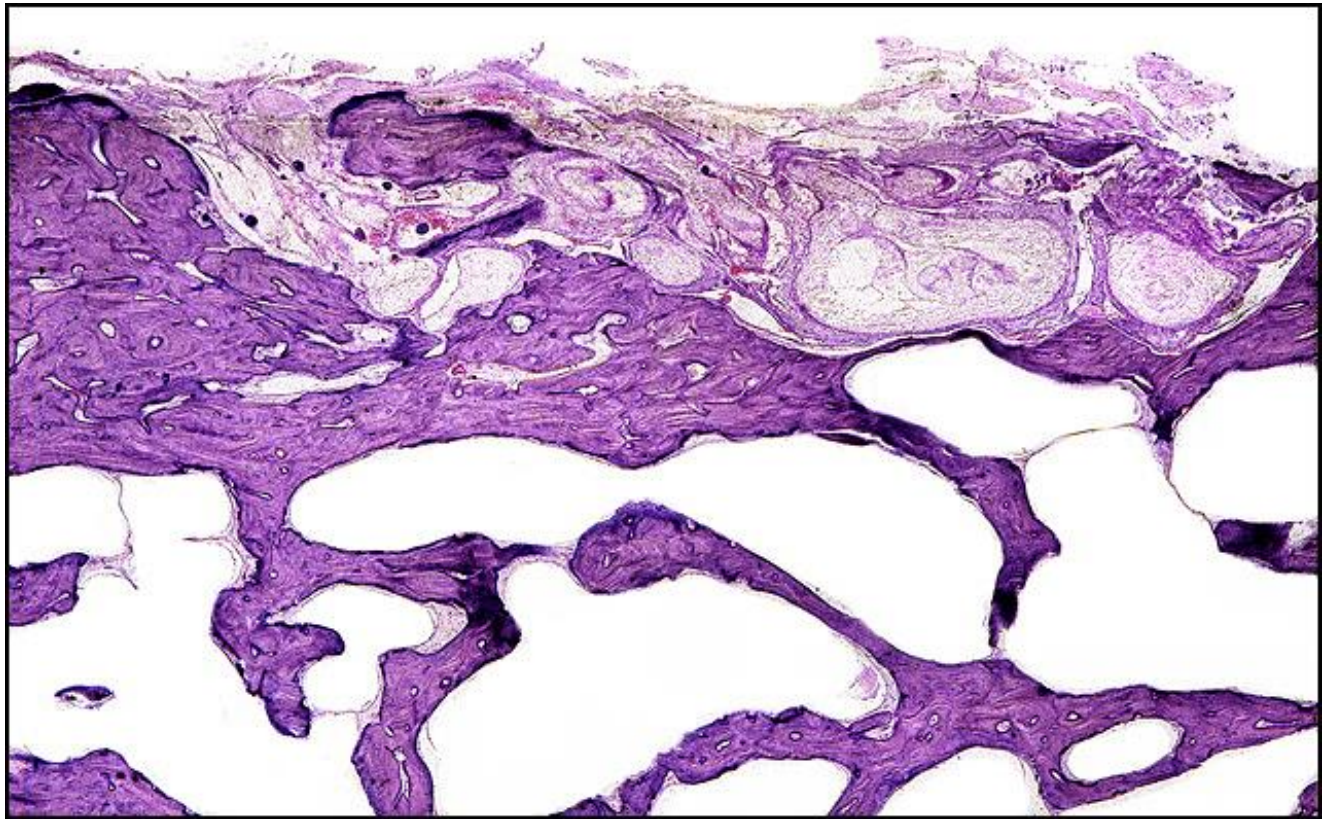
TB Case Number: 117

Gender: Female

Age (yrs.): 90

Otologic Diagnosis:

1. Paget's disease, osteoblastic and remodeled phase, mild, left
2. Internal auditory canal, narrowed, Paget's disease, without nerve compression, left
3. Pacchionian bodies, mastoid, left
4. Artifact, preparation, membranous labyrinth, moderately severe



Arachnoid Granulation, R-161

Title: Arachnoid Granulation, R-161

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

TB Case Number: 66

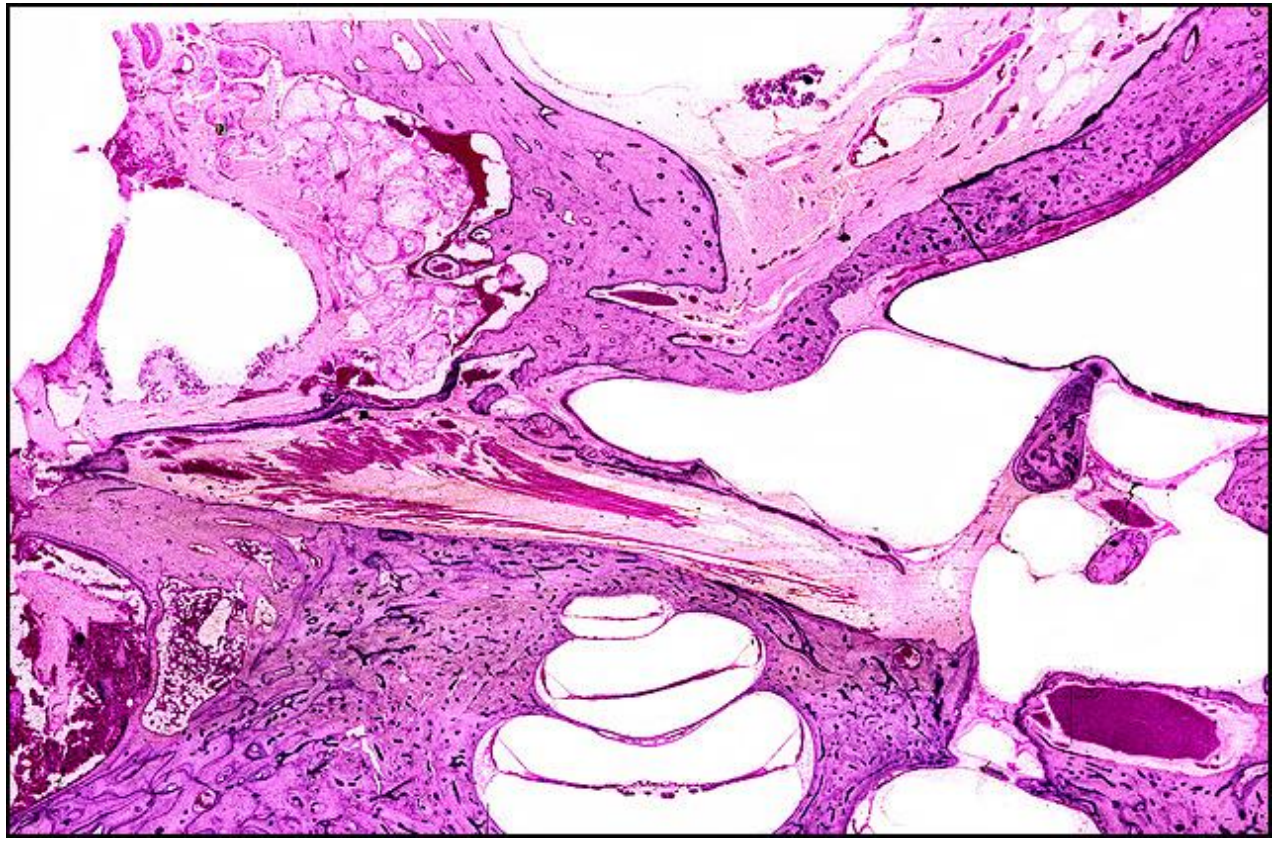
Comments: Arachnoid granulation, R-161

Gender: Male

Age (yrs.): 51

Otologic Diagnosis:

1. Temporal bones, normal
2. Membranous labyrinth, normal, bilateral
3. Artery, accessory meningeal, bilateral
4. Facial hiatus, widely open, bilateral
5. Artifact, preparation, compression, right
6. Pacchionian body, floor of middle cranial fossa, right



Arachnoid Granulation, R-161

Title: Arachnoid Granulation, R-161

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

TB Case Number: 66

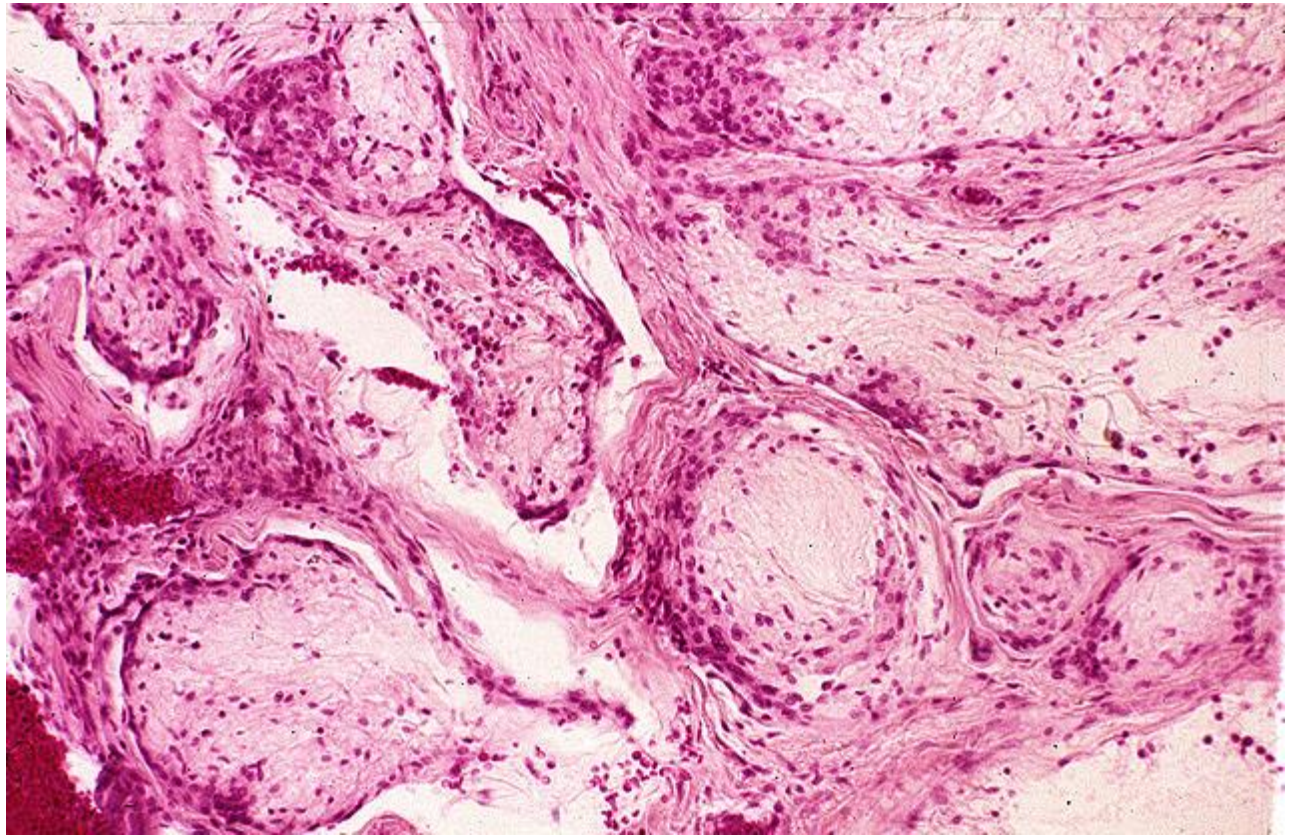
Comments: Arachnoid granulations

Gender: Male

Age (yrs.): 51

Otologic Diagnosis:

1. Temporal bones, normal
2. Membranous labyrinth, normal, bilateral
3. Artery, accessory meningeal, bilateral
4. Facial hiatus, widely open, bilateral
5. Artifact, preparation, compression, right
6. Pacchionian body, floor of middle cranial fossa, right



Arachnoid Granulation, L-100

Title: Arachnoid Granulation, L-100

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

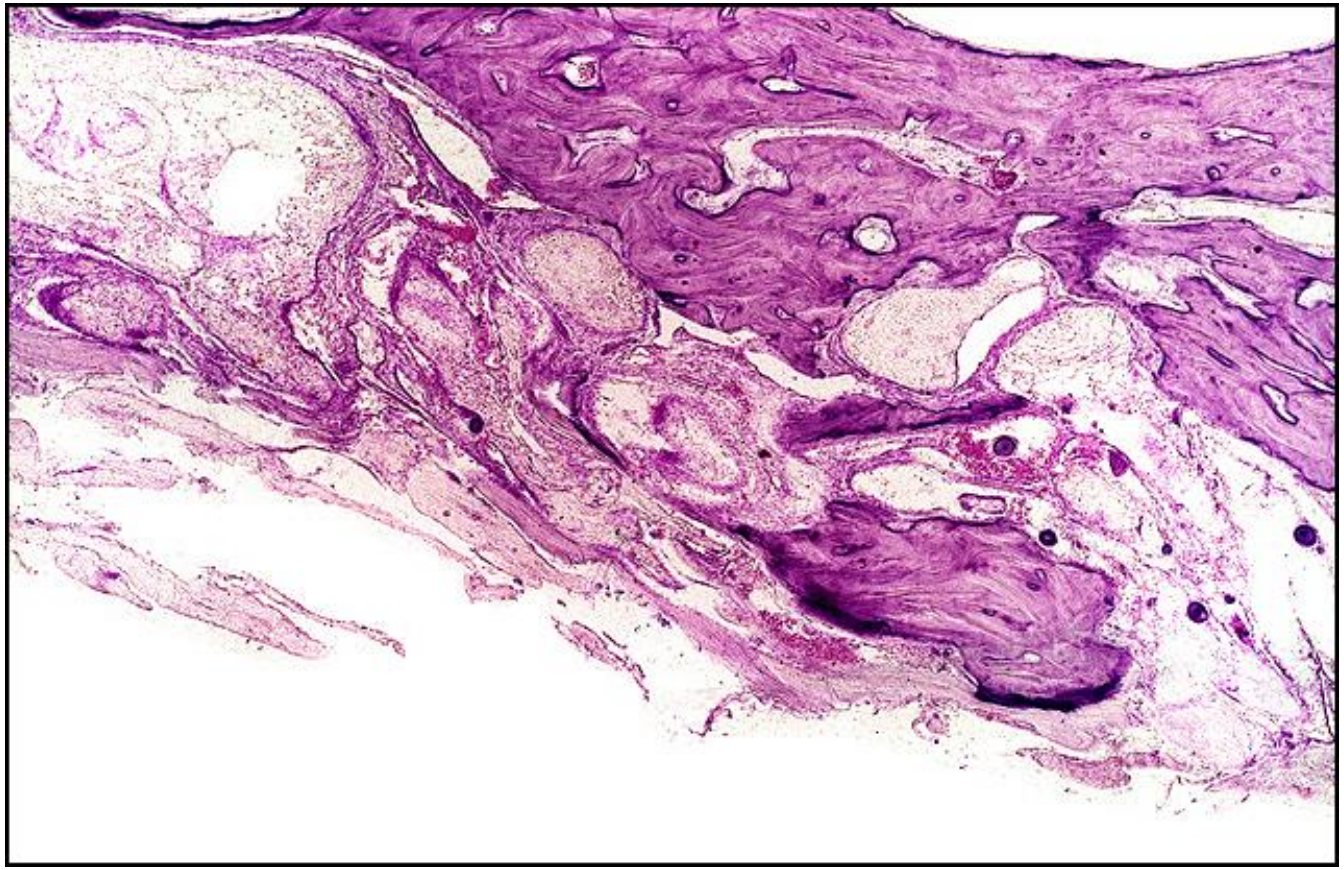
TB Case Number: 117

Gender: Female

Age (yrs.): 90

Otologic Diagnosis:

1. Paget's disease, osteoblastic and remodeled phase, mild, left
2. Internal auditory canal, narrowed, Paget's disease, without nerve compression, left
3. Pacchionian bodies, mastoid, left
4. Artifact, preparation, membranous labyrinth, moderately severe



Tympanomeningeal Fissure, L-401

Title: Tympanomeningeal Fissure, L-401

Chapter: Developmental Defects

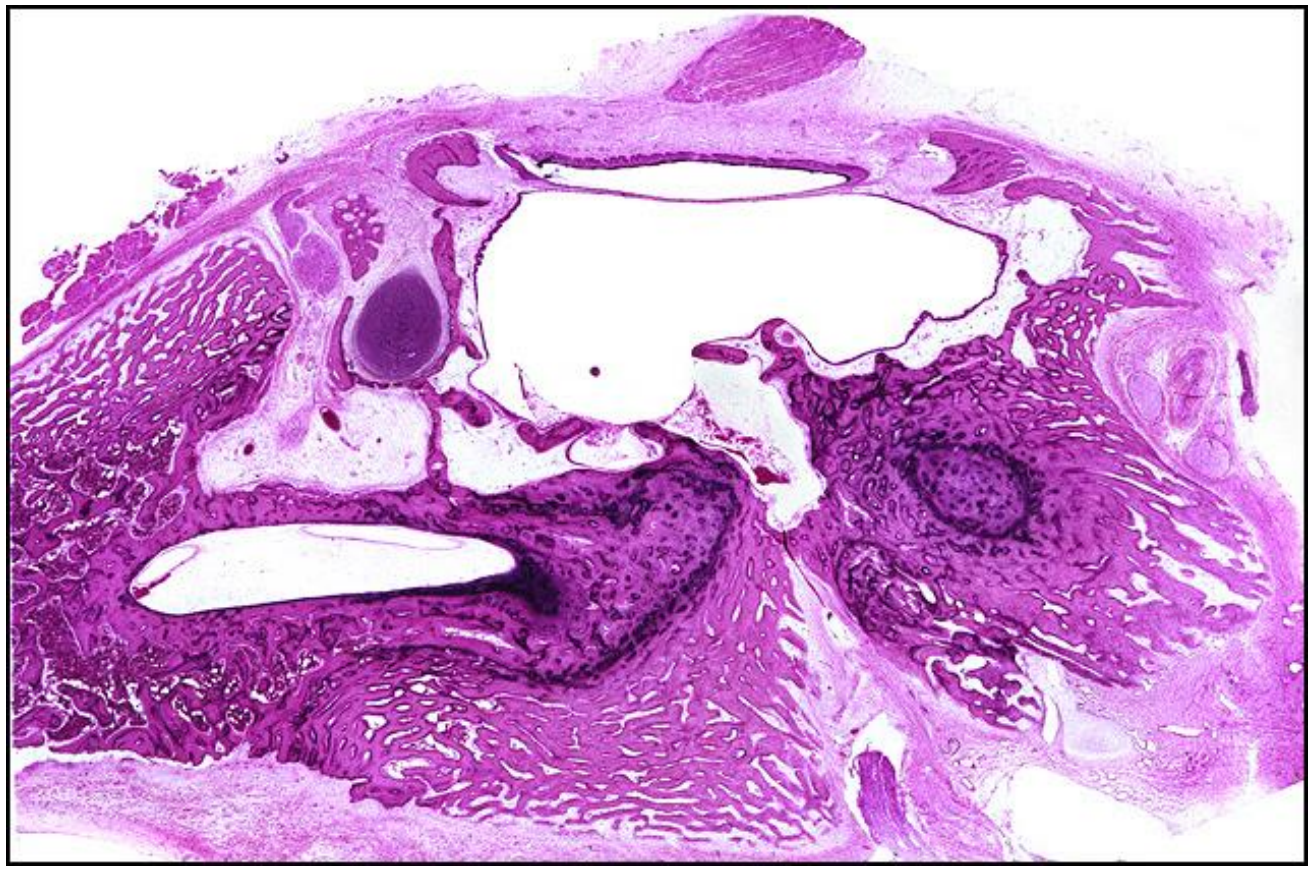
Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

TB Case Number: 145

Gender: Female

Otologic Diagnosis:

1. Temporal bone, newborn, normal, left
2. Membranous labyrinth, normal, left
3. Bony labyrinth, normal, left
4. Hemorrhage, recent, perilymphatic space, cochlear and vestibular labyrinths, left
5. Styloid eminence, normal, left
6. Tympanomeningeal



Peritubal Air Cell, R-401

Title: Peritubal Air Cell, R-401

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

TB Case Number: 1024

Comments: 94 yr old female showing a peritubal air cell that opens into the bony eustachian tube, anterior to the level of the tympanic membrane

Gender: Female

Age (yrs.): 95

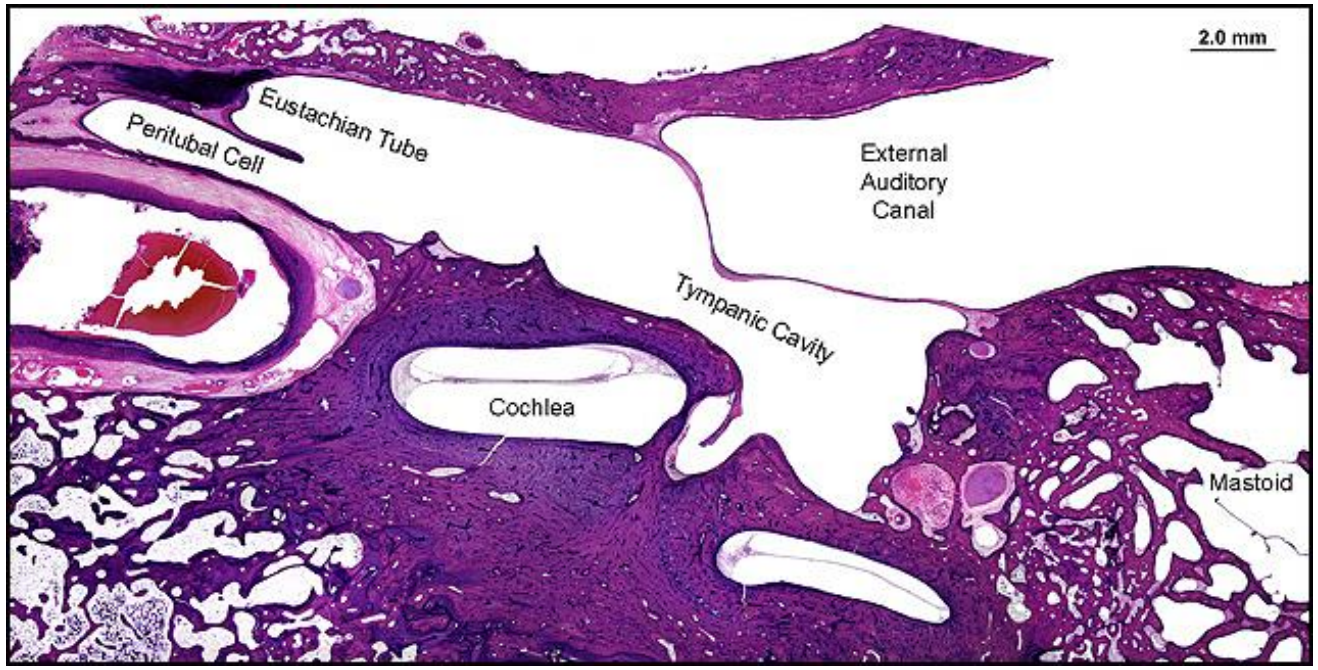
Otologic Diagnosis:

Clinical:

Adult onset, progressive, bilateral sensorineural hearing loss, probably genetic in etiology

Histopathologic:

Stria vascularis atrophy, near total, affecting entire cochlear duct, bilateral, probably genetically determined



Peritubal Air Cell, R-381.2

Title: Peritubal Air Cell, R-381.2

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

TB Case Number: 1005

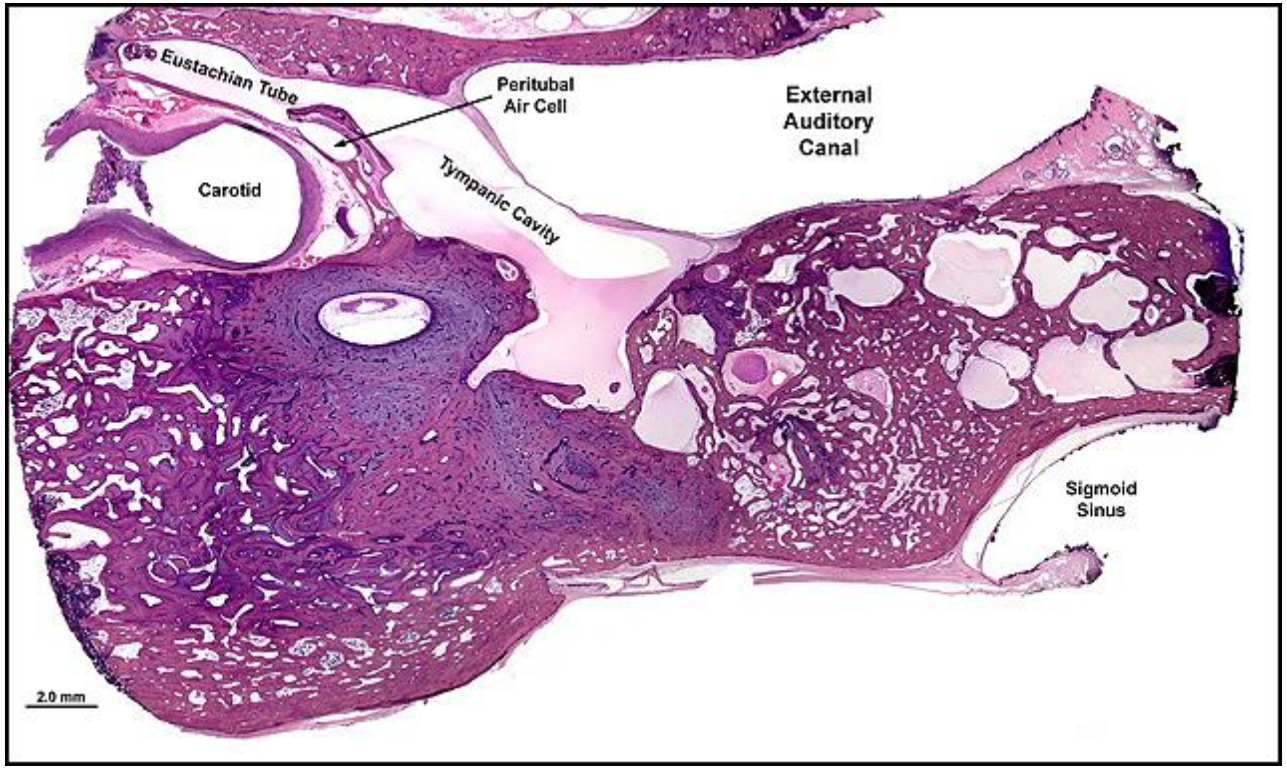
Comments: Showing opening of peritubal air cell into ET, anterior to tympanic membrane, 92-year-old female.

Gender: Female

Age (yrs.): 92

Otologic Diagnosis:

Mixed hearing loss, AU, predominantly sensorineural, due to presbycusis



Occult Meningo-Encephalocele, R-81

Title: Occult Meningo-Encephalocele, R-81

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

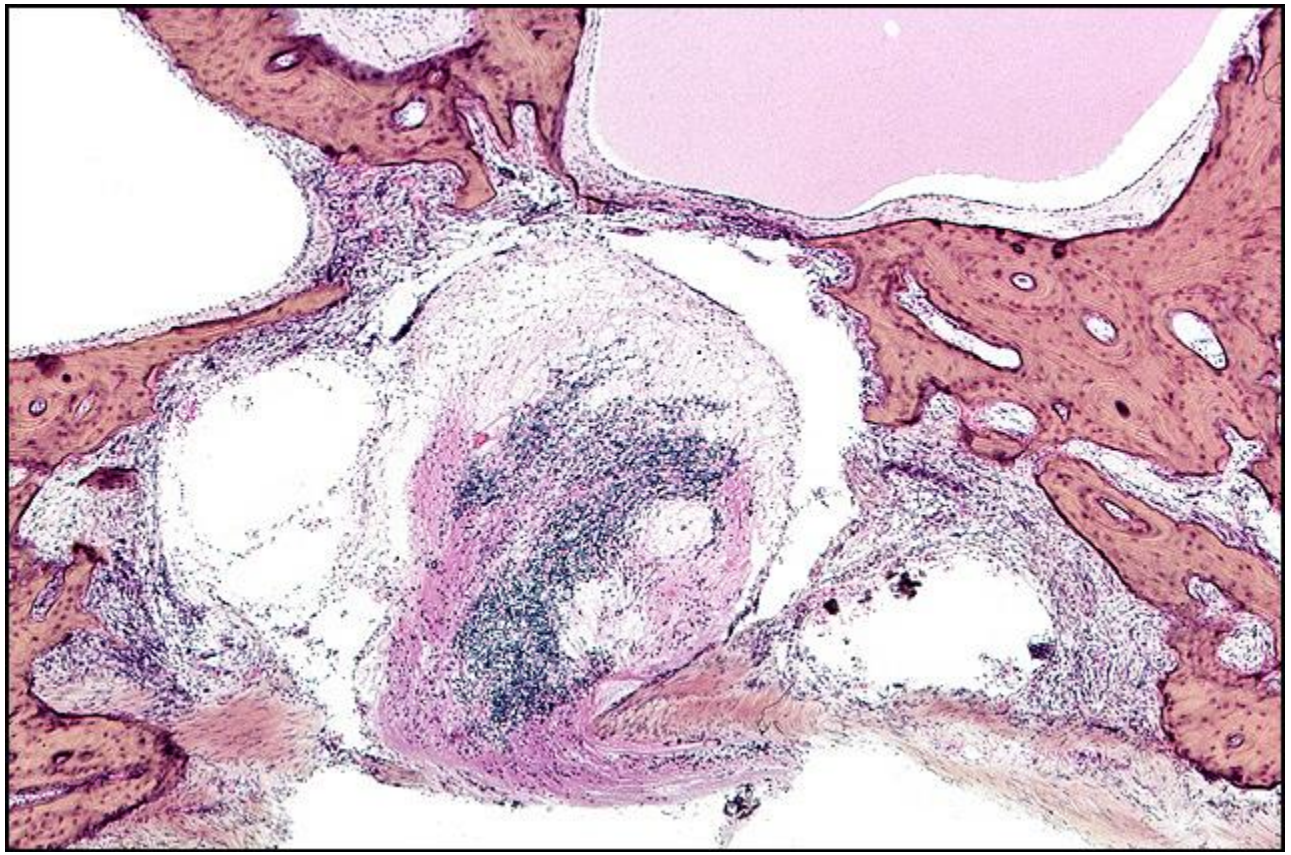
TB Case Number: 939

Gender: Male

Age (yrs.): 75

Otologic Diagnosis:

1. Cochleovestibular schwannoma, clinical, right
2. Endolymphatic hydrops, secondary to #1, right
3. Degeneration of hair cells and spiral ganglion cells, secondary to #1, right
4. Eosinophilic staining of endolymphatic and perilymphatic spaces



Occult Meningo-Encephalocele, R-81

Title: Occult Meningo-Encephalocele, R-81

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

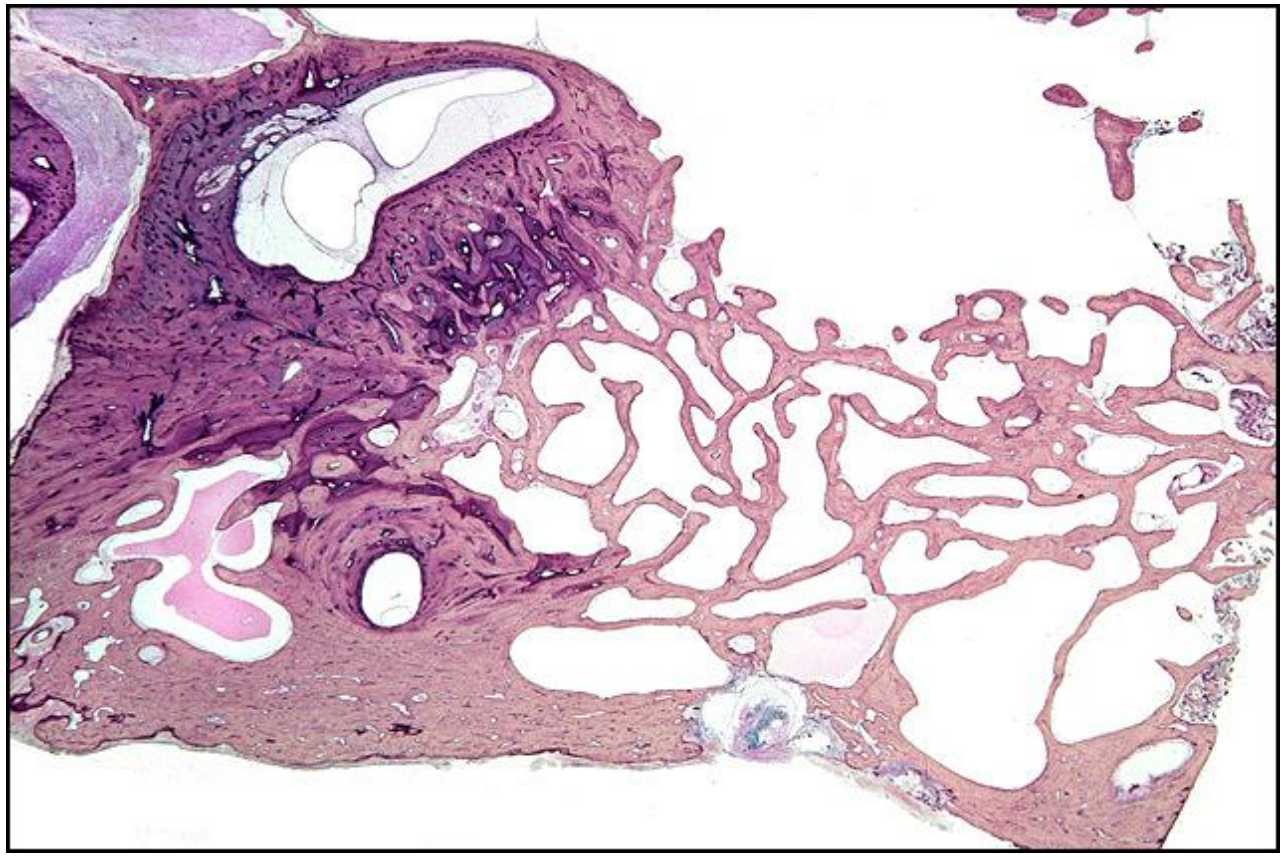
TB Case Number: 939

Gender: Male

Age (yrs.): 75

Otologic Diagnosis:

1. Cochleovestibular schwannoma, clinical, right
2. Endolymphatic hydrops, secondary to #1, right
3. Degeneration of hair cells and spiral ganglion cells, secondary to #1, right
4. Eosinophilic staining of endolymphatic and perilymphatic spaces



CSF Leak, R-251

Title: CSF Leak, R-251

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

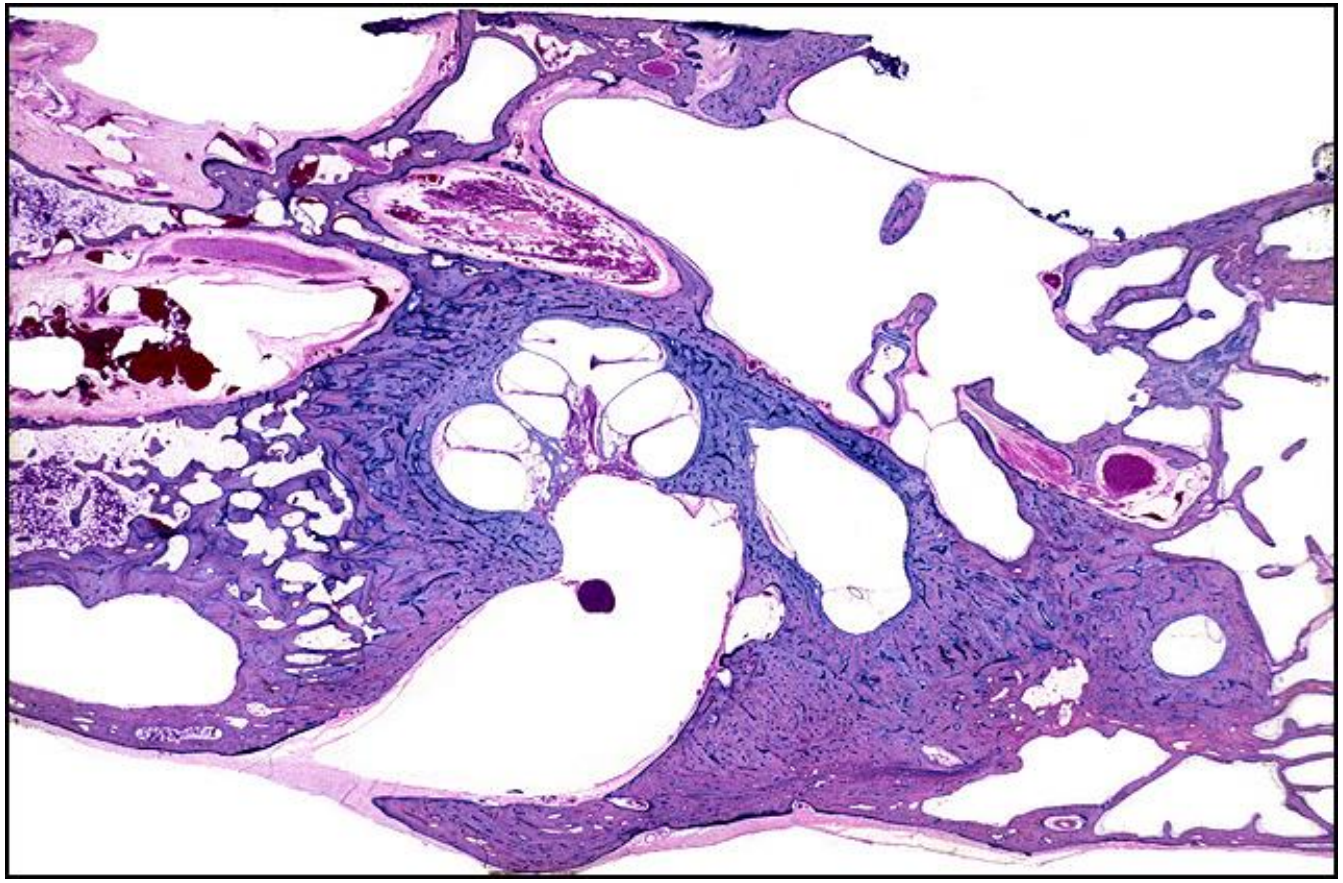
TB Case Number: 113

Gender: Male

Age (yrs.): 71

Otologic Diagnosis:

1. Cerebrospinal fluid (CSF) otorrhea, post traumatic, delayed, left
2. Meningitis, suppurative, complicating CSF otorrhea
3. Abscess, brain, otogenic, temporal lobe, left
4. Fracture, temporal bone, tegmen, tympanic, presumptive, left
5. Myringotomy



CSF Leak, L-141

Title: CSF Leak, L-141

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

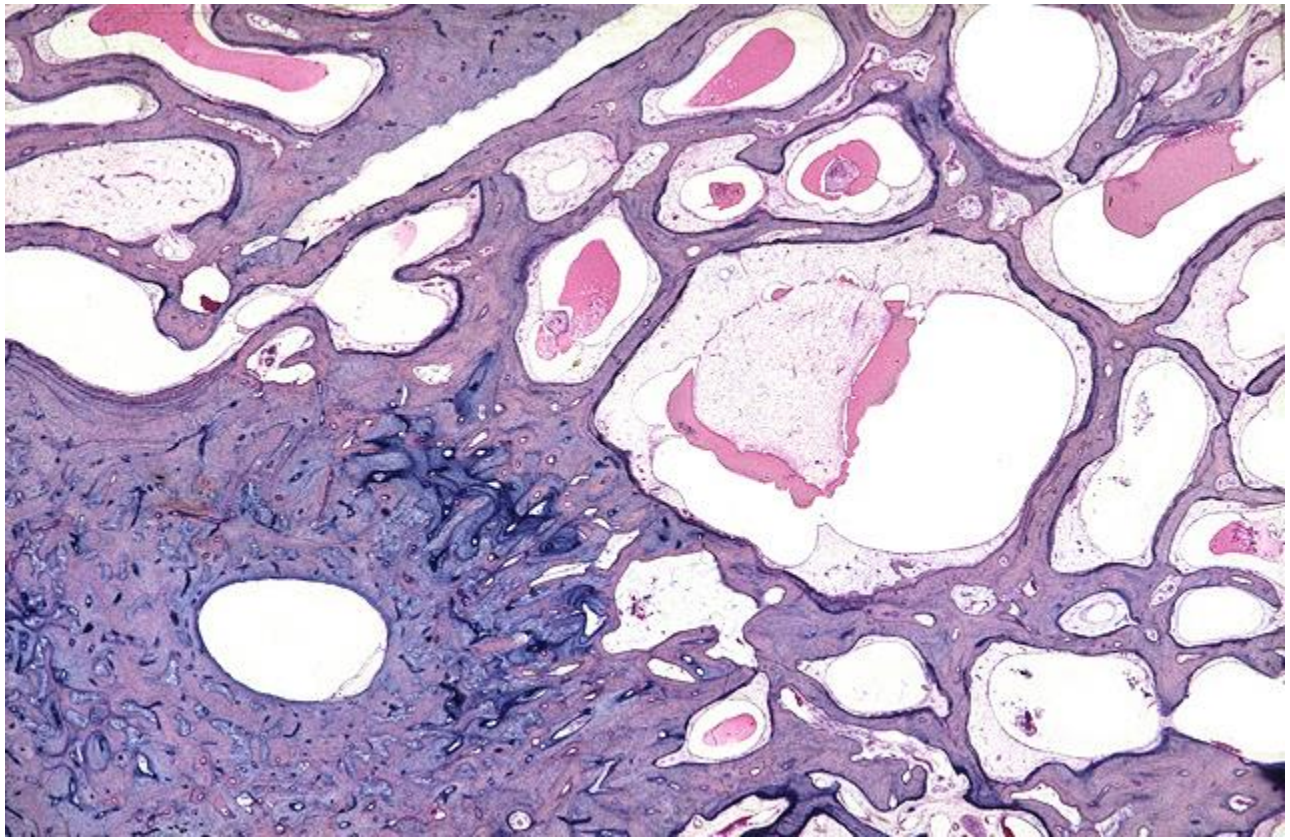
TB Case Number: 113

Gender: Male

Age (yrs.): 71

Otologic Diagnosis:

1. Cerebrospinal fluid (CSF) otorrhea, post traumatic, delayed, left
2. Meningitis, suppurative, complicating CSF otorrhea
3. Abscess, brain, otogenic, temporal lobe, left
4. Fracture, temporal bone, tegmen, tympanic, presumptive, left
5. Myringotomy



CSF Leak, L-460

Title: CSF Leak, L-460

Chapter: Developmental Defects

Chapter Section: CSF Leaks, Dural Defects, Variform Anomalies External and Middle Ear

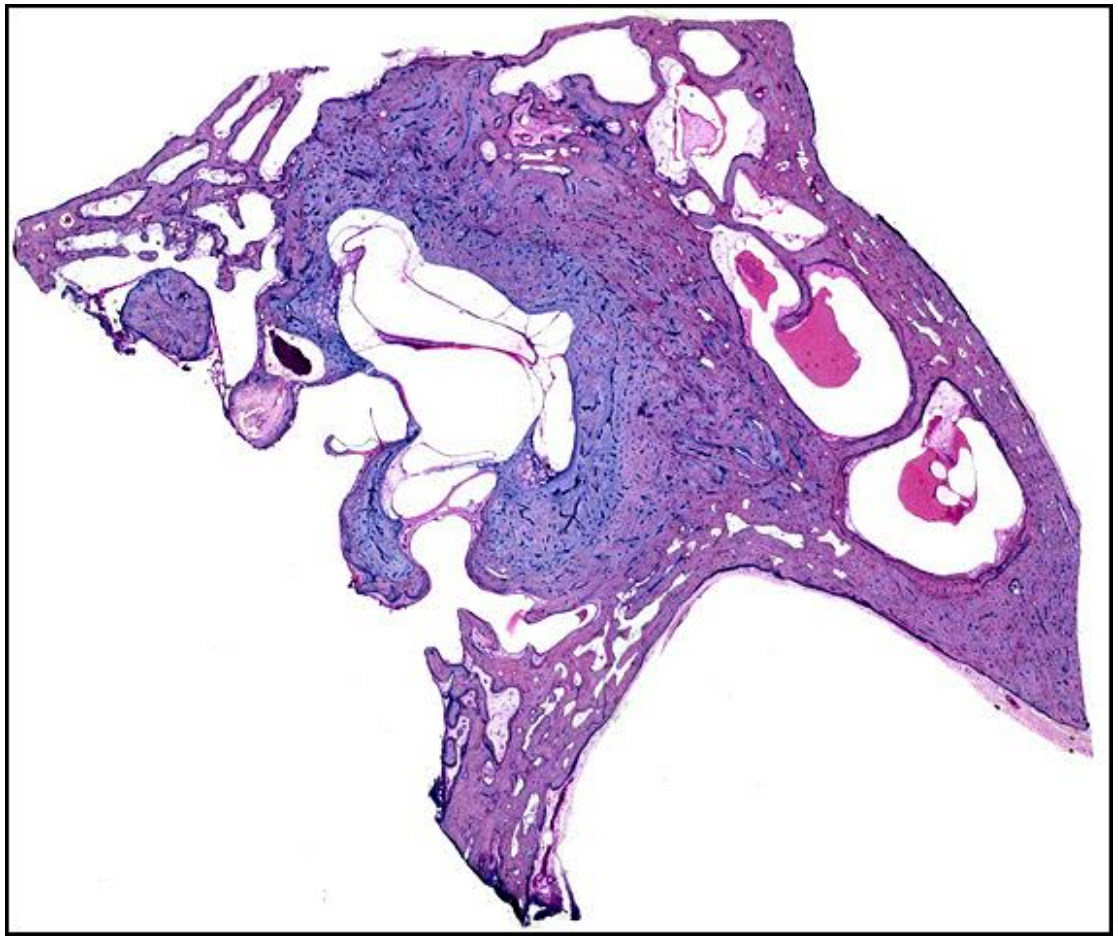
TB Case Number: 113

Gender: Male

Age (yrs.): 71

Otologic Diagnosis:

1. Cerebrospinal fluid (CSF) otorrhea, post traumatic, delayed, left
2. Meningitis, suppurative, complicating CSF otorrhea
3. Abscess, brain, otogenic, temporal lobe, left
4. Fracture, temporal bone, tegmen, tympanic, presumptive, left
5. Myringotomy



Cochlear Nerve Agenesis, R-141

Title: Cochlear Nerve Agenesis, R-141

Chapter: Developmental Defects

Chapter Section: Cochlear Anomalies, Inner Ear, Variform Anomalies

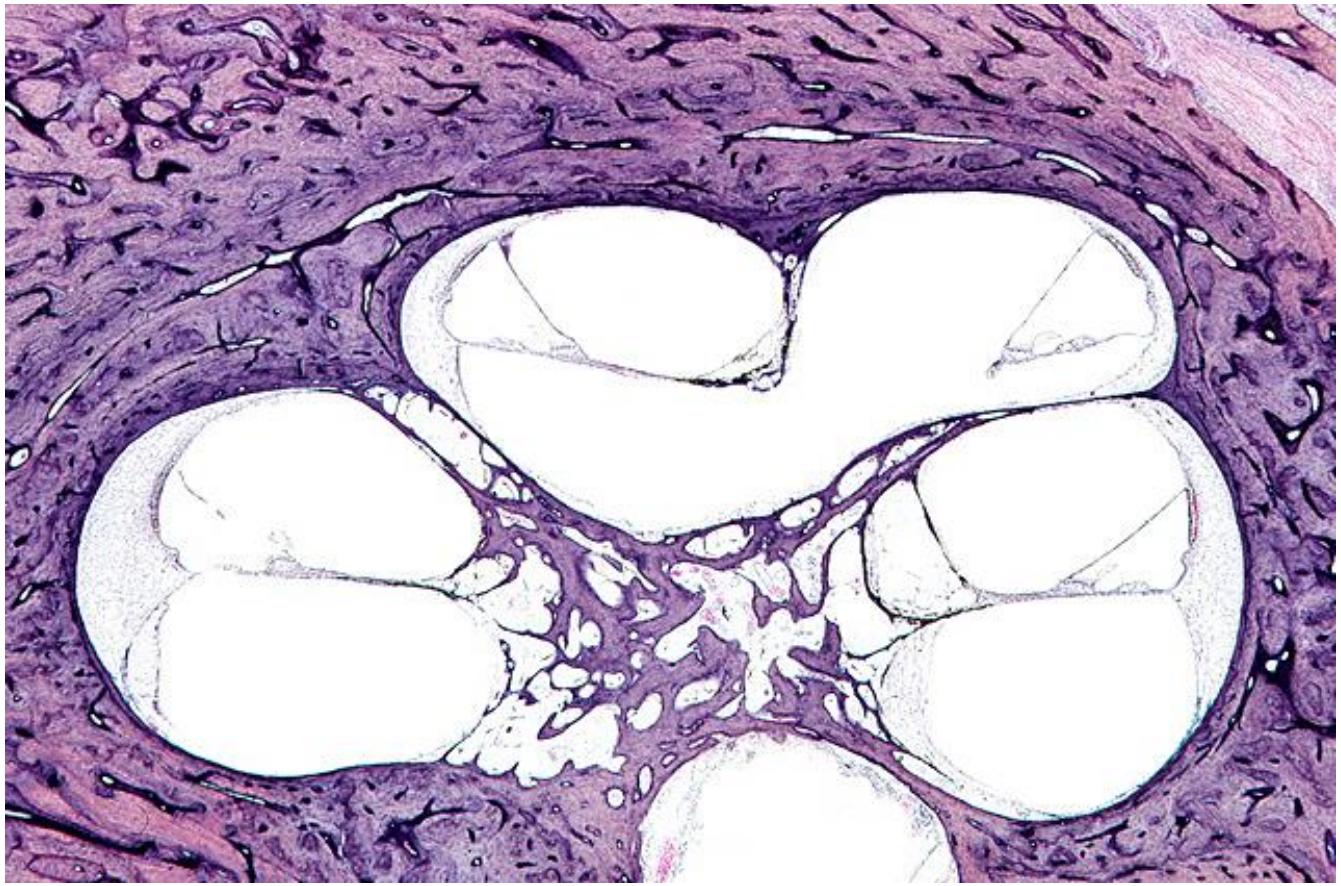
TB Case Number: 935

Gender: Male

Age (yrs.): 10

Otologic Diagnosis:

1. Mondini dysplasia, bilateral
2. Agenesis of cochlear nerve, bilateral
3. Acute and chronic otitis media, bilateral
4. Tympanic membrane, central perforation, left
5. Tympanosclerosis involving tympanic membrane and mesotympanum, left
6. Jugular bulb



Cochlear Nerve Agenesis, R-141.2

Title: Cochlear Nerve Agenesis, R-141.2

Chapter: Developmental Defects

Chapter Section: Cochlear Anomalies, Inner Ear, Variform Anomalies

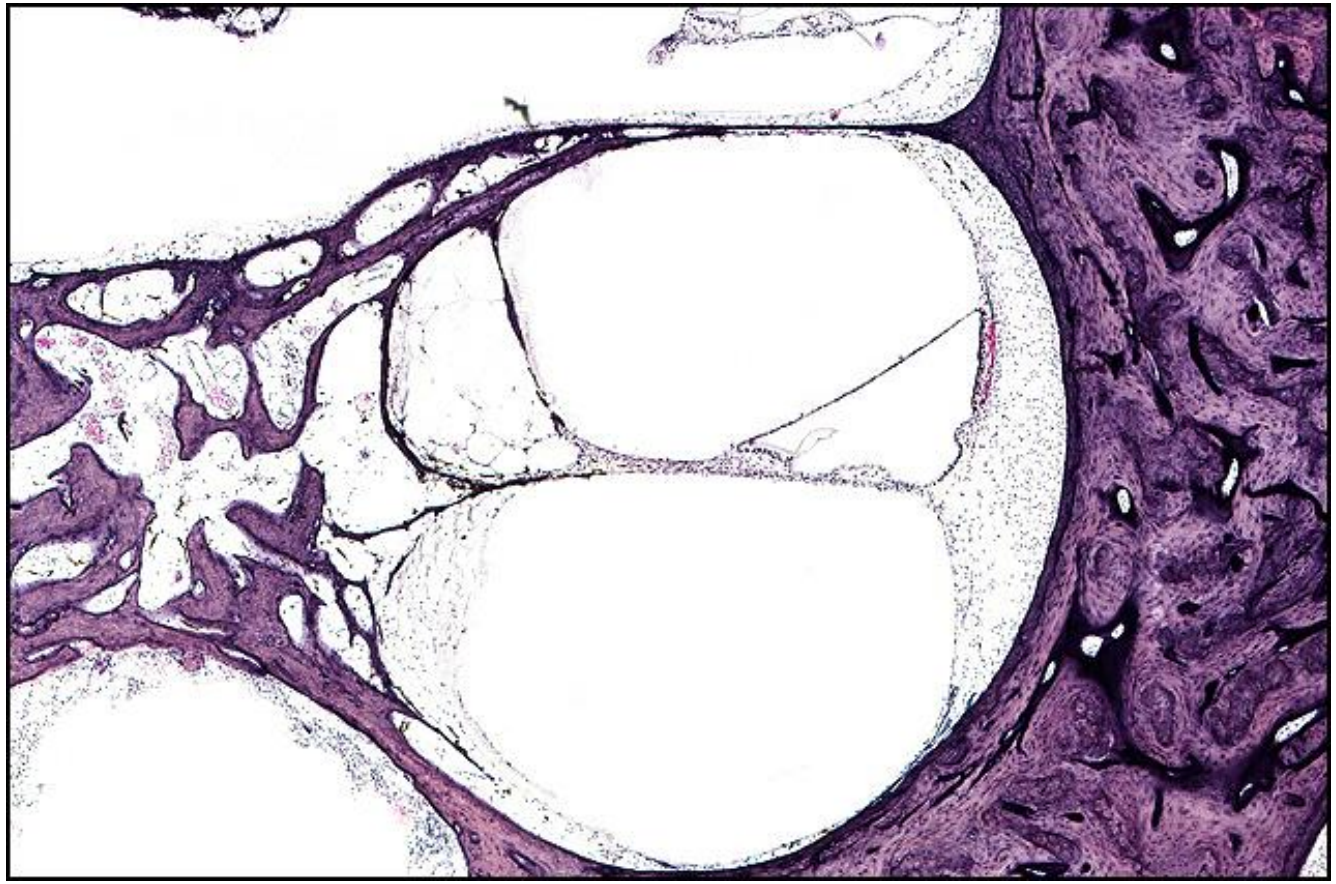
TB Case Number: 935

Gender: Male

Age (yrs.): 10

Otologic Diagnosis:

1. Mondini dysplasia, bilateral
2. Agenesis of cochlear nerve, bilateral
3. Acute and chronic otitis media, bilateral
4. Tympanic membrane, central perforation, left
5. Tympanosclerosis involving tympanic membrane and mesotympanum, left
6. Jugular bulb



Cochlear Nerve Agenesis, R-171

Title: Cochlear Nerve Agenesis, R-171

Chapter: Developmental Defects

Chapter Section: Cochlear Anomalies, Inner Ear, Variform Anomalies

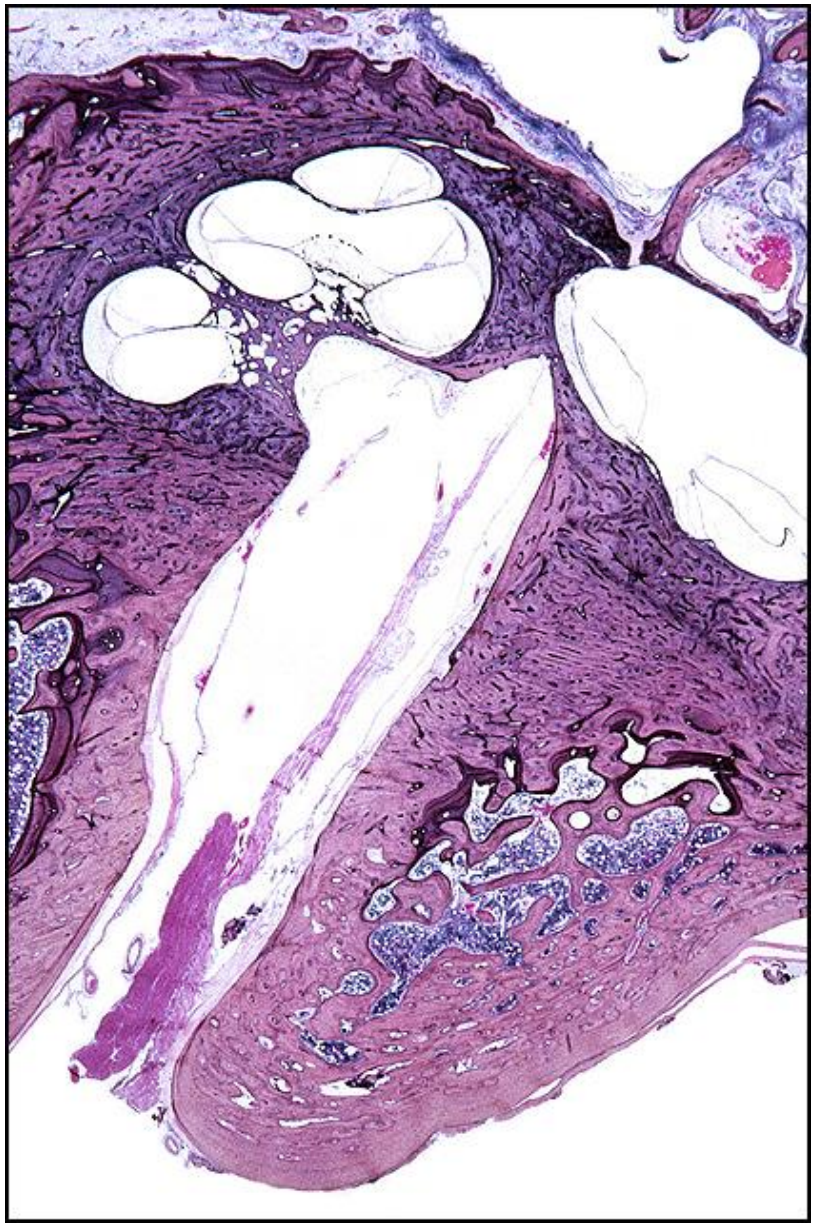
TB Case Number: 935

Gender: Male

Age (yrs.): 10

Otologic Diagnosis:

1. Mondini dysplasia, bilateral
2. Agenesis of cochlear nerve, bilateral
3. Acute and chronic otitis media, bilateral
4. Tympanic membrane, central perforation, left
5. Tympanosclerosis involving tympanic membrane and mesotympanum, left
6. Jugular bulb



Absent UE Valve, L-208

Title: Absent UE Valve, L-208

Chapter: Developmental Defects

Chapter Section: Inner Ear, Variform Anomalies, Vestibular Labyrinth Anomalies

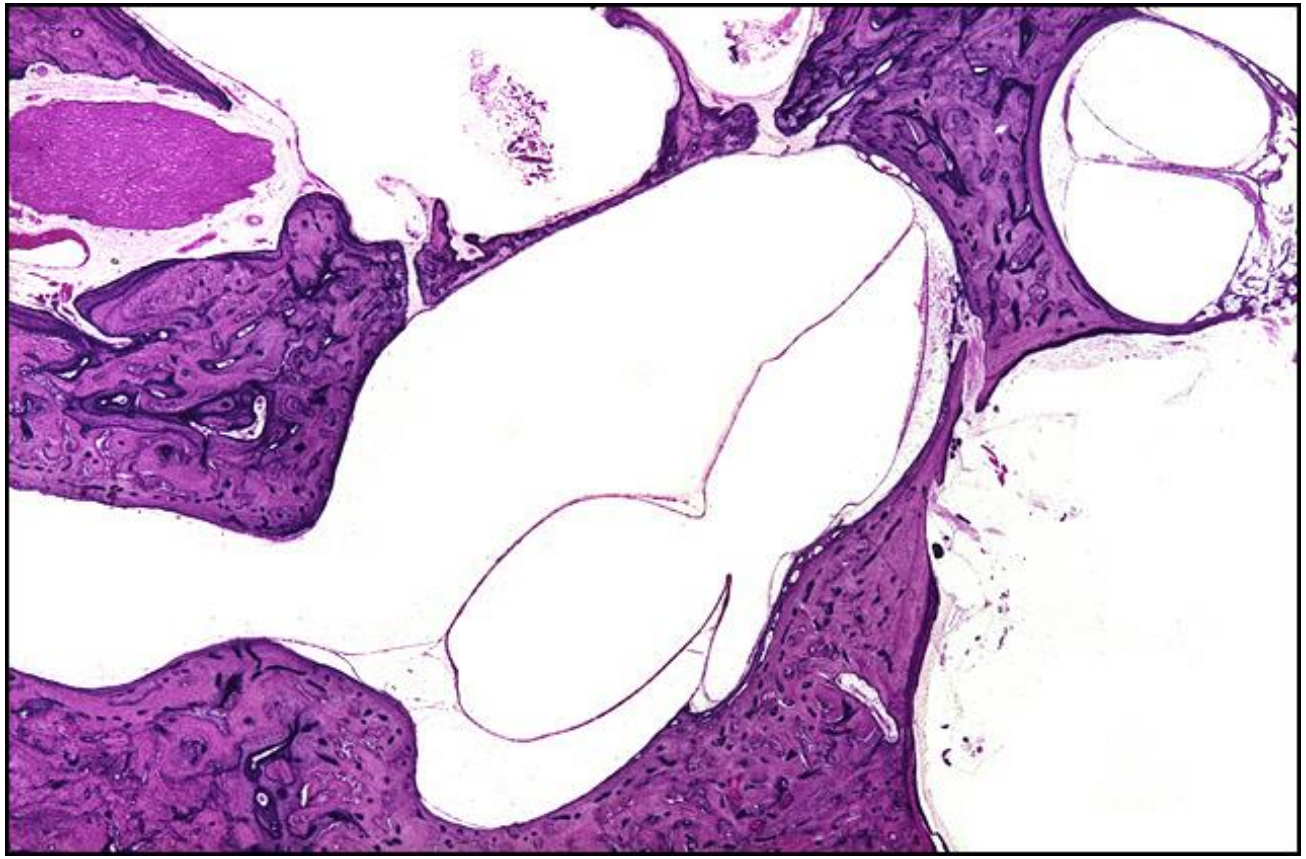
TB Case Number: 473

Gender: Male

Age (yrs.): 55

Otologic Diagnosis:

1. Streptomycin, ototoxicity
2. Anomaly, saccule, bilateral
    a) Utriculoendolymphatic valves, absent
    b) Utricles and saccules in wide communication with each other and with endolymphatic sinuses
3. Artifact



Enlarged Vestibular Aqueduct, R-391

Title: Enlarged Vestibular Aqueduct, R-391

Chapter: Developmental Defects

Chapter Section: Enlarged Vestibular Aqueduct, Inner Ear, Variform Anomalies

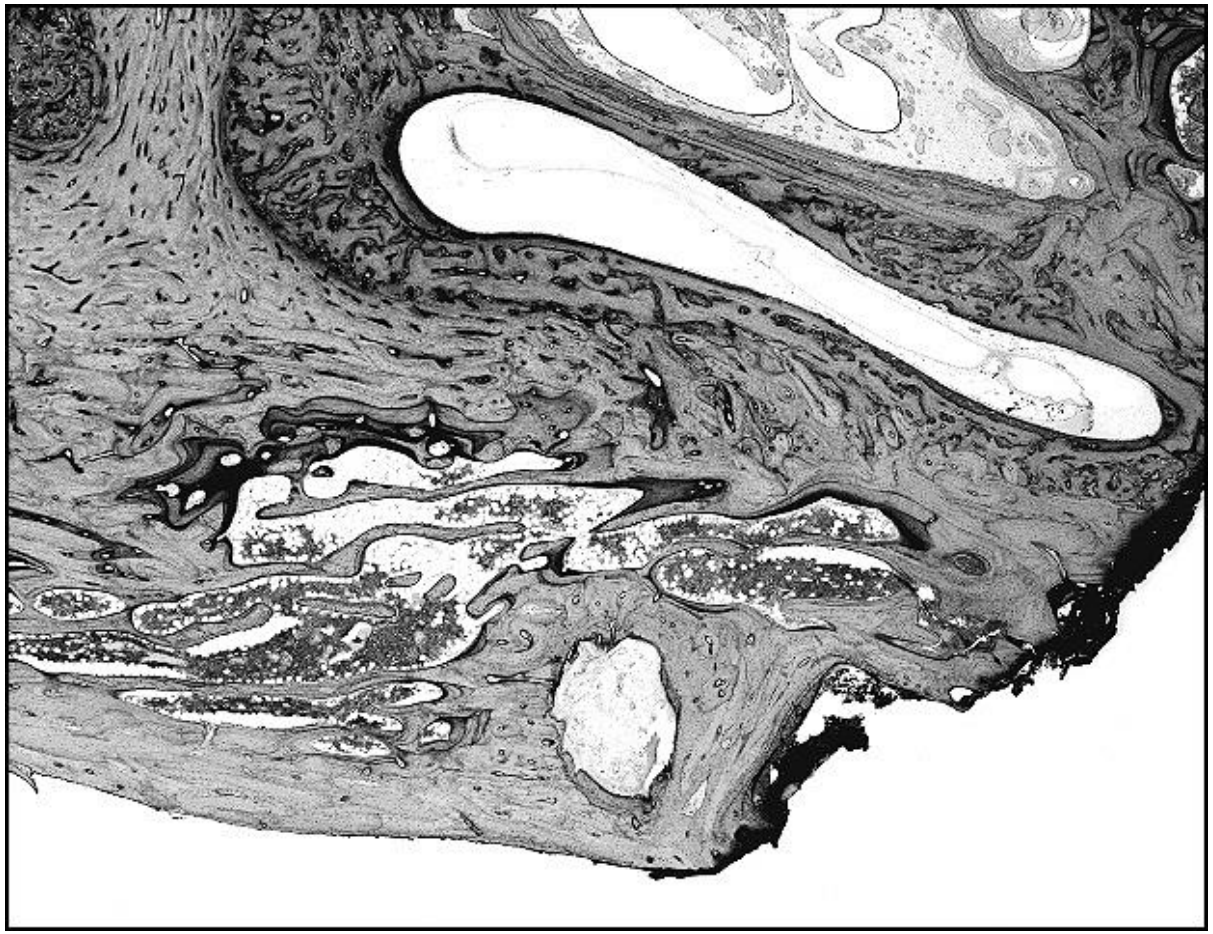
TB Case Number: 904

Gender: Male

Age (yrs.): 6

Otologic Diagnosis:

1. Otitis media, active, chronic
2. Mastoiditis, active, chronic
3. Effusion, middle ear and mastoid, chronic
4. Incus, anomaly, ankylosis
5. Anomaly, cochlea, mild, enlarged Scala tympani in basal and middle turns
6. Saccule, endolymphatic distension/hydrops



Enlarged Vestibular Aqueduct, R-251

Title: Enlarged Vestibular Aqueduct, R-251

Chapter: Developmental Defects

Chapter Section: Enlarged Vestibular Aqueduct, Inner Ear, Variform Anomalies

TB Case Number: 904

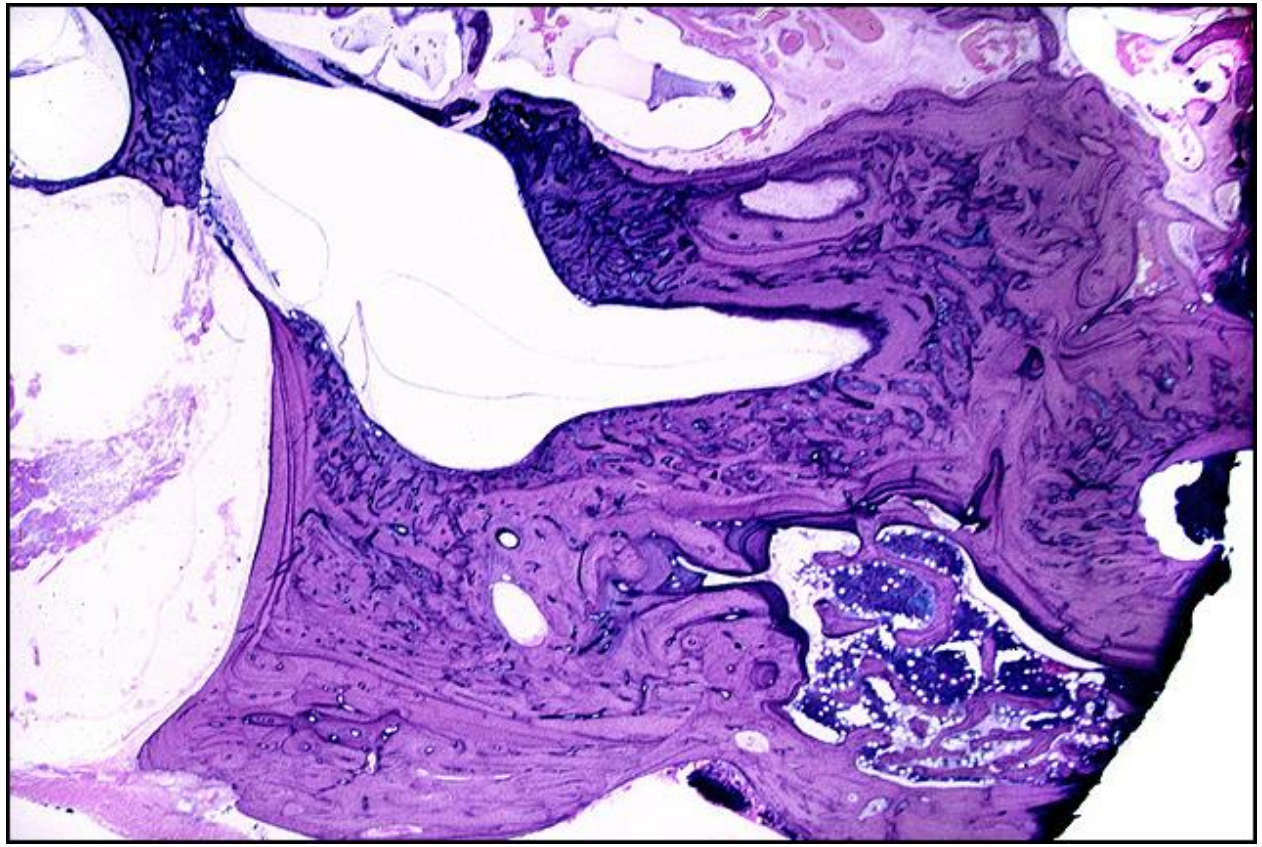
Comments: Also, Dubowitz Syndrome

Gender: Male

Age (yrs.): 6

Otologic Diagnosis:

1. Otitis media, active, chronic
2. Mastoiditis, active, chronic
3. Effusion, middle ear and mastoid, chronic
4. Incus, anomaly, ankylosis
5. Anomaly, cochlea, mild, enlarged Scala tympani in basal and middle turns
6. Saccule, endolymphatic distension/hydrops



Dubowitz Syndrome, R-221

Title: Dubowitz Syndrome, R-221

Chapter: Developmental Defects

Chapter Section: Enlarged Vestibular Aqueduct, Inner Ear, Variform Anomalies

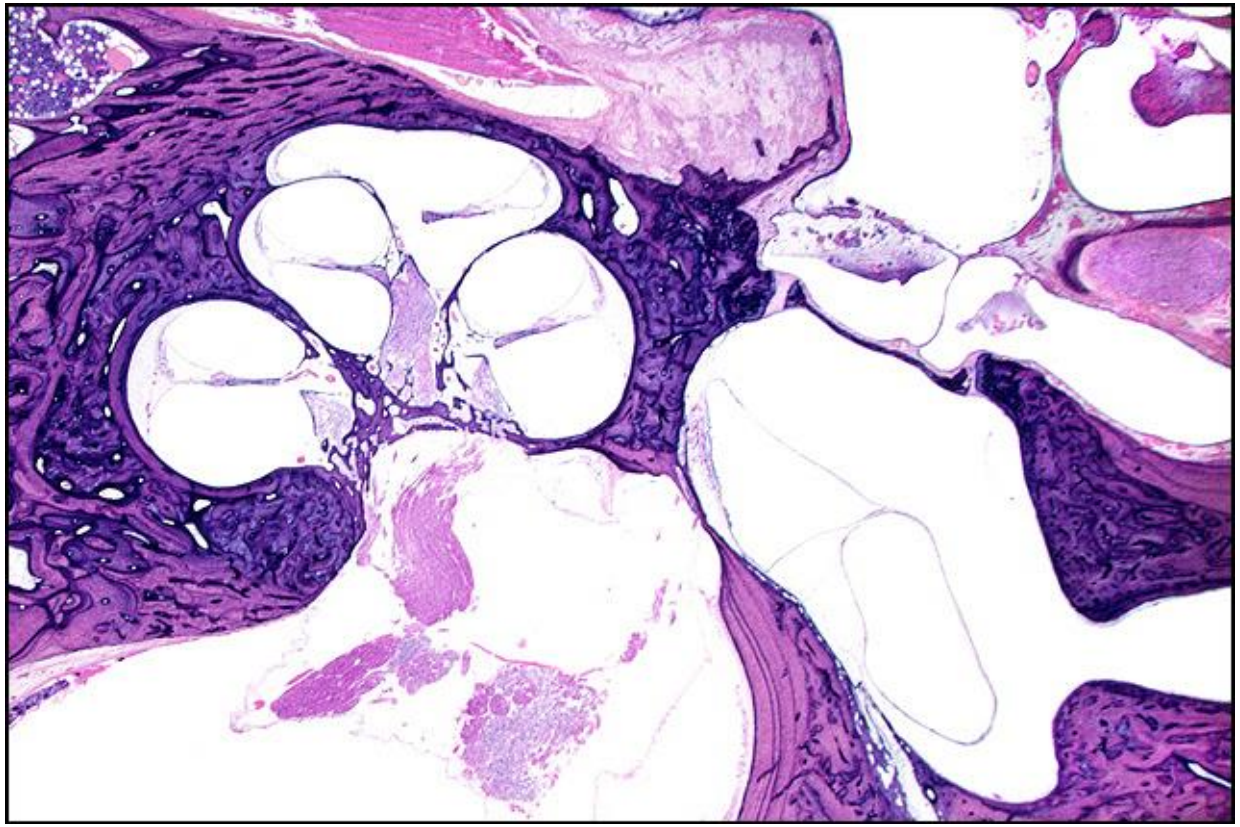
TB Case Number: 904

Gender: Male

Age (yrs.): 6

Otologic Diagnosis:

1. Otitis media, active, chronic
2. Mastoiditis, active, chronic
3. Effusion, middle ear and mastoid, chronic
4. Incus, anomaly, ankylosis
5. Anomaly, cochlea, mild, enlarged Scala tympani in basal and middle turns
6. Saccule, endolymphatic distension/hydrops



EVA with Bony Septum, R-341

Title: EVA with Bony Septum, R-341

Chapter: Developmental Defects

Chapter Section: Enlarged Vestibular Aqueduct, Inner Ear, Variform Anomalies

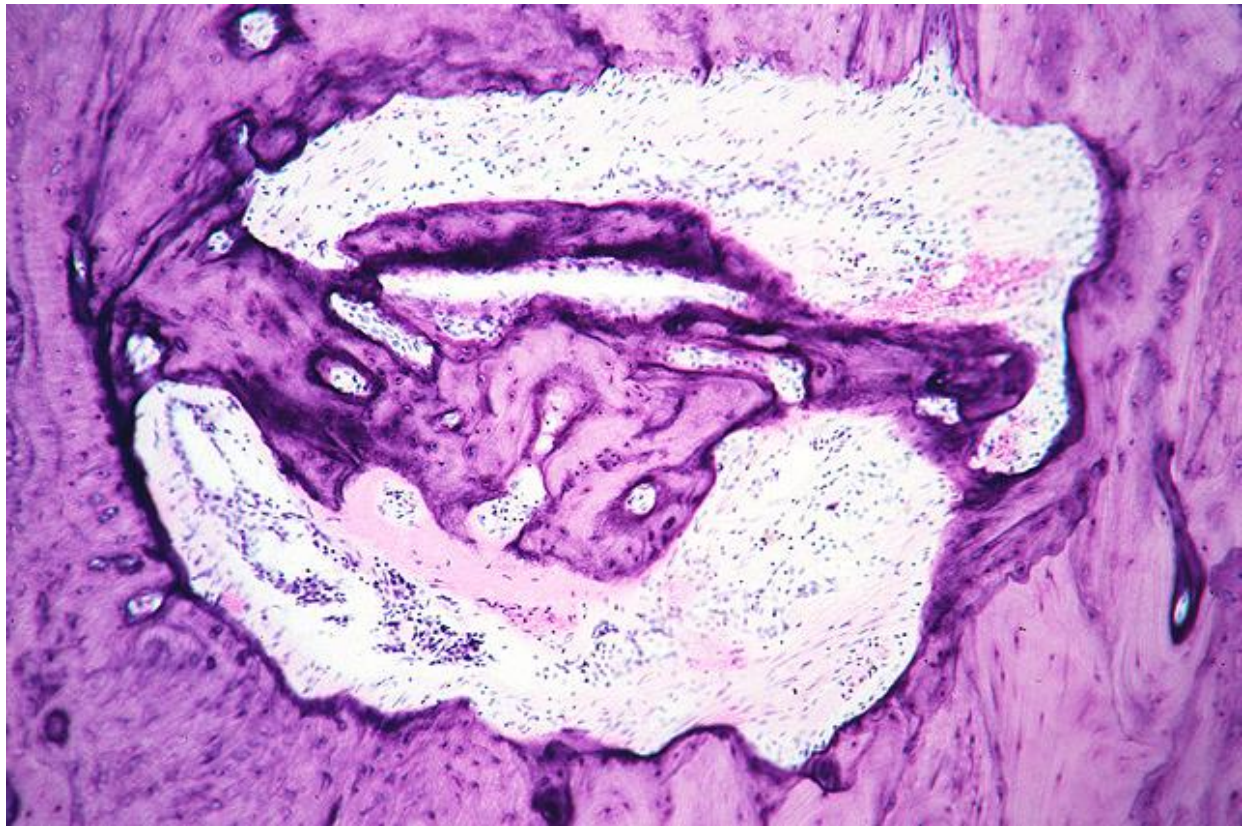
TB Case Number: 904

Comments: Enlarged vestibular aqueduct, also Dubowitz Syndrome

Age (yrs.): 6

Otologic Diagnosis:

1. Otitis media, active, chronic
2. Mastoiditis, active, chronic
3. Effusion, middle ear and mastoid, chronic
4. Incus, anomaly, ankylosis
5. Anomaly, cochlea, mild, enlarged Scala tympani in basal and middle turns
6. Saccule, endolymphatic distension/hydrops



Mass General Brigham

Mass Eye and Ear