Syndromes which may feature accessory tragus: an overview

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Accessory tragus is an easily recognized congenital cutaneous anomaly. There are syndromes, which feature accessory tragi. These are Delleman syndrome, Goldenhar syndrome, Haberland syndrome, and Townes-Brock syndrome. However, these syndromes, which are named eponymously, are also known in the medical literature by other names. The Online Mendelian Inheritance of Man (OMIM), which is well-known data base for genetic diseases and syndromes, also, lists them by different names. Table 1 sheds some lights on these uncommon syndromes.

These syndromes differ in their prevalence but may share few clinical features. For instance, colobomas occur in all of these syndromes along with a variety of neurologic abnormalities. Definitive diagnosis of each of the above syndromes is often challenging owing to phenotypic variability and overlap with other congenital syndromes. However, there are some differentiating features which help in reaching the right diagnosis for each of them. For example; aplasia cutis, lack of auricular abnormalities, and the presence of neurologic abnormalities usually distinguish Delleman syndrome from the closely related Goldenhar syndrome.

<table>
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<tr>
<th>The syndrome</th>
<th>OMIM number</th>
<th>Remarks</th>
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<tbody>
<tr>
<td>Delleman syndrome</td>
<td>164180</td>
<td>Named in OMIM, as &quot;Oculocerebrocutaneous syndrome&quot;. Also known as,&quot;Orbital cyst with cerebral and focal dermal malformations&quot;. First described by Delleman and Oorthuys, in 1981. It is characterized by ocular, brain, and skin anomalies.</td>
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<td>Goldenhar syndrome</td>
<td>164210</td>
<td>Named in OMIM as &quot;Hemifacial microsomia&quot;. Also known as, &quot;Oculoauriculovertebral syndrome or spectrum&quot;. First described by Maurice Goldenhar, in 1952. The classic triad associated with the syndrome includes the presence of limbal dermoids, preauricular skin tags and pretragal fistulas.</td>
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<td>Haberland syndrome</td>
<td>613001</td>
<td>Named in OMIM, as &quot;Encephalocraniocutaneous lipomatosis&quot;. First described by Haberland and Perou, in 1970. It is an example of ectomesodermal dysgenesis characterized by profound mental retardation, early onset of seizures, unilateral temporofrontal lipomatosis, ipsilateral cerebral and leptomeningeal lipomatosis, cerebral malformation and calcification, and lipomas of the skull, eye, and heart.</td>
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<tr>
<td>Townes-Brocks syndrome</td>
<td>107480</td>
<td>Named in OMIM, as &quot;Townes-Brocks syndrome&quot;. Also, known as Renal–Ear–Anal–Radial syndrome or REAR syndrome. First described by Townes and Brocks, in 1972. It is a well-described genetic syndrome characterized by anal, ear, and thumb anomalies</td>
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When a newborn presents with multiple accessory tragi, the patient should be carefully examined for the presence of other congenital anomalies. Early recognition of a potential congenital syndrome is important to direct appropriate evaluation and management and provide anticipatory guidance to families.\(^1\)

References