

# Elongated styloid process (Eagle's syndrome)

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**Introduction:** The styloid process shows lot of variations in its length. In majority of patients it is about 20 – 30 mm long. Technically speaking when the length of styloid process exceeds 30 mm then it is considered to be elongated. The clinical signs and symptoms associated with elongated styloid process was first described by Eagle in 1937. Later this condition became known as Eagle's syndrome / Elongated styloid process. The signs and symptoms of elongated styloid process are pretty vague and often at best misleading. These patients usually go medical shopping visiting neurologists, dental surgeons, psychiatrists and surgeons. The diagnosis of this condition requires awareness and vigilance. This condition can be confirmed by palpating the tonsillar fossa, infiltration of local anesthetic agents and imaging studies.

**History:**

Historically the ossification of stylohyoid apparatus can be divided into three periods. This division is purely for better understanding. Era of anatomists: Anatomists belonging to 17th century described ossification of stylohyoid apparatus they encountered during dissection as normal variants as they were not privy to the clinical details and patient history. Era of diagnostic radiologists: This period includes the early 20th century. Due to advances in radiological anatomy, radiologists were able to identify ossification of stylohyoid apparatus and correlate this condition with that of the symptoms expressed by the patient. Eagle under whom this syndrome is named belonged to this era. Era of panoramic radiology: This period includes the mid 20th century. Routine study of panoramic radiographs by dental surgeons threw up more such cases of ossification of the stylohyoid apparatus.

**Anatomy:** Embryologically the styloid process is derived from the second branchial arch ( a component of Reichert's cartilage). It is a slender bony structure extending antero inferiorly from the petrosal aspect of temporal bone. In front of the styloid process the following structures are seen:

1. Internal maxillary artery
2. Lingual nerve

### 3. Auriculotemporal nerves

Posterior to the styloid process the following structures are seen:

1. Internal jugular vein
2. Internal carotid artery
3. Cervical sympathetic chain
4. Last 4 cranial nerves (9,10,11, and 12)

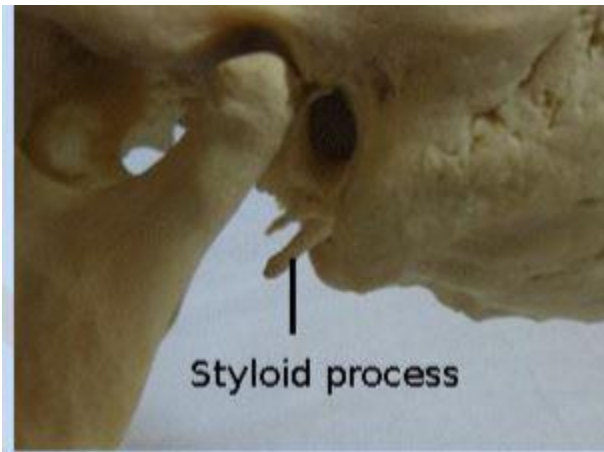


Figure showing the styloid process

Structures attaching to the styloid process:

These include:

1. Stylopharyngeus muscle – medially
2. Stylohyoid muscle – laterally
3. Styloglossus muscle – anteriorly
4. Two ligaments stylohyoid and stylomandibular also gets attached to this process

Gossman's classification of types of elongated styloid processes: Gossman studies about 4000 patients with elongated styloid process and classified it into three types.

1. Elongated
2. Crooked
3. Segmented
4. Very elongated

Correll's classification of elongated styloid process:

Type I: Elongated styloid process

Type II: Pseudoarticulated styloid process

Type III: Segmental styloid process

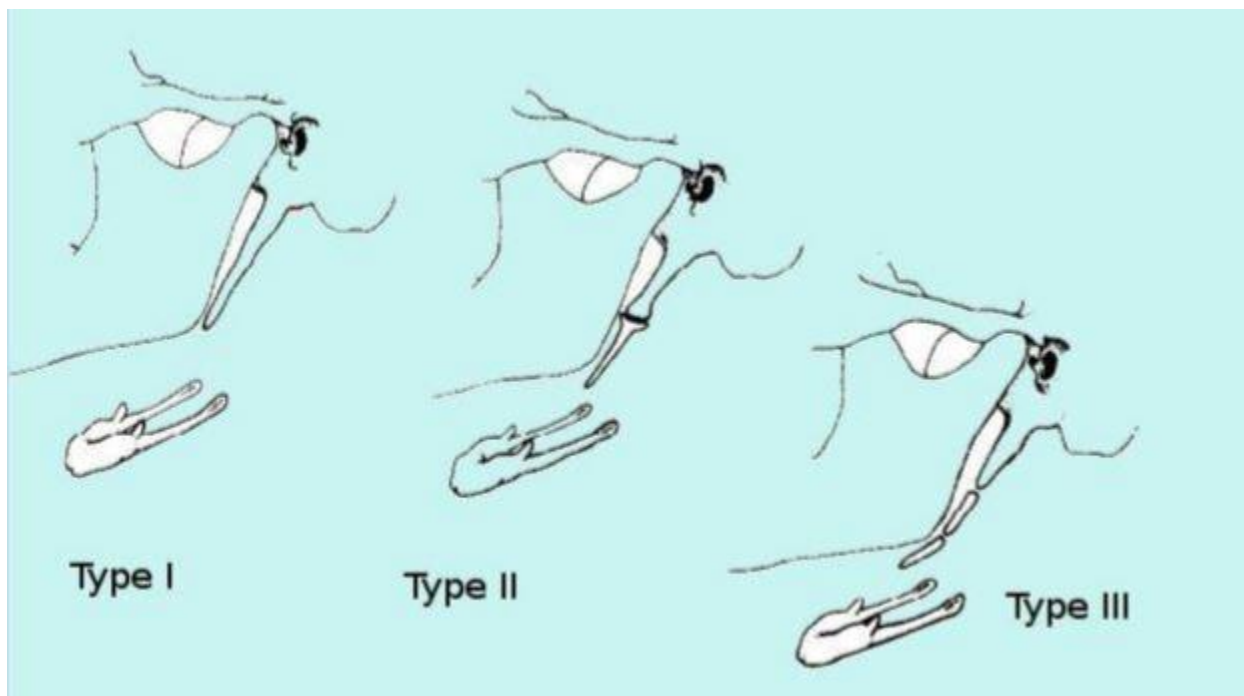


Figure showing the types of styloid process as described by Correll

Langlais classification:

This classification suggested by Robert Langlais included the three types as described by Correll, to facilitate radiological classification of elongated styloid process included the term calcification. He describes 4 types of calcifications in addition to the three types of styloid process as described by Correll.

<b>Correll's classification</b>	<b>Calcification pattern of styloid process</b>
Type I - Elongated	Calcified outline
Type II – Pseudo articulated	Partially calcified
Type III - Segmental	Nodular
	Completely calcified

Type I elongated styloid process: Radiologically this type of styloid process appears as an uninterrupted image, its length ranging from 25 – 30 mm. Radiographically a styloid process which is 25 mm long is considered to be elongated styloid process. If orthopantomograms are studied a styloid process of about 28 cm is considered to be normal because of the inherent magnification involved in this imaging modality.

Type II (Pseudoarticulated variety): In this type the styloid process is joined to the mineralized stylomandibular / stylohyoid ligament through a single pseudo articulation. This articulation commonly appears superior to the level of the inferior border of the mandible. Radiologically this type of styloid process appears like an articulated styloid process.

Type III (segmental variety): This type is composed of non continuous portions of styloid process due to interruptions in the mineralized segments. Radiologically it appears like segmental mineralized stylohyoid complex.

Patterns of calcification seen in elongated styloid process:

1. Calcified outline: Is seen in a majority of elongated styloid process. Radiologically it appears with a thin radio opaque border with central lucency (resembling radiographs of long bones).
2. Partially calcified stylohyoid process: Radiologically this type of styloid process has a thicker radio opaque outline with almost complete opacification in some areas.

3. Nodular complex: This type of styloid process has a knobby / scalloped outline, with partial or complete calcification

4. Complete calcification: This type of styloid process appears radiologically as completely calcified with no radiolucent inner core.



Symptoms: Common symptoms associated with elongated styloid process include:

1. Vague pain in the neck
2. Foreign body sensation in the throat
3. Pain in the throat
4. Painful swallowing
5. Pain while changing head position
6. Pain in the ear
7. Pain over temporomandibular joint
8. Pain radiating to upper limb

Probable causes of stylalgia:

1. Fracture of ossified stylohyoid ligament – caused by trauma, sudden laughter or epileptic seizures
2. Nerve compression by elongated / malpositioned styloid process. Glossopharyngeal nerve is commonly involved
3. Degenerative and inflammatory changes associated with elongated styloid process
4. Irritation of pharyngeal mucosa
5. Impingement of carotid vessels by the elongated styloid process (carotidynia)

Classic features of stylalgia: include

1. Dull and nagging pain

2. Pain becomes worse on deglutition 3. Pain radiates to the ear and mastoid region

Note: Eagle's syndrome should be considered in all patients with vague craniofacial pain.

Eagle classically described two types of Symptom complexes.

Classic Eagle's syndrome: Commonly develops in patients following tonsillectomy. These patients have persistent throat pain and globus pallidus. These symptoms could be caused due to contraction of post tonsillectomy scar tissue towards the elongated styloid process resulting in the impingement of one or more of the following cranial nerves i.e. 5,7,9 and 10.

Carotid artery syndrome: In this type the carotid arteries are intermittently compressed during head turning movements of neck. Head rotation in these patients classically causes compression of internal carotid artery and sympathetic chain resulting in syncope, ipsilateral headache and orbital pain. Compression of external carotid artery causes pain in the distribution of temporal and maxillary branches.

Clinical tests to confirm elongated styloid process:

1. Palpation of tonsillar fossa: This elicits similar pain / aggravation of pre existing pain. 2. Xylocaine infiltration test: Patients suspected of having elongated styloid process on being infiltrated about 2 ml of 2% lignocaine into the tonsillar fossa have significant reduction in pain. A positive xylocaine infiltration test usually indicates Eagle's syndrome

Theories of ossification of stylohyoid apparatus:

In humans the ceratohyal element of second branchial arch degenerates with time. It should be noted that its fibrous sheath, which has a potential to ossify persists as stylohyoid ligament. The stylohyoid process ossifies between 5-8 years after birth, and any variation in this ossification process leads to the creation of elongated styloid process. Hence the term ossification should be ideally used instead of calcification.

Steinmann's theory of ossification of styloid apparatus:

Steinmann proposed three theories to account for ossification of styloid apparatus.

**Theory of reactive hyperplasia:** This theory suggests that if the styloid process is appropriately stimulated its terminal end undergoes ossification at the expense of stylohyoid ligament. The stimulus could even be pharyngeal trauma.

**Theory of reactive metaplasia:** This theory suggests that traumatic stimulus would induce certain ligamentous sections of stylohyoid ligament to undergo metaplastic changes provoking intermittent ossification of the same. Metaplasia is possible due to the presence of osseous centres within the stylohyoid ligament. When stimulated these osseous centres becomes ossified forming osseous links causing ossification of stylohyoid ligament.

**Theory of anatomic variance:** This theory suggests that the stylohyoid process and stylohyoid ligament gets ossified very early in life. This phenomenon could be considered as normal anatomical variant. This theory accounts for the presence of elongated styloid process in childhood.

According to Steinmann true Eagle's syndrome may either be caused by reactive hyperplasia or reactive metaplasia of stylohyoid apparatus. This does not include the symptom complex caused by long standing ossified stylohyoid complex as is the case in the theory of anatomical variance. These patients should ideally managed conservatively. Occasionally elderly patients may present with symptoms of Eagle's syndrome without radiological evidence of elongated styloid process. These symptoms may be explained by the theory of aging and developmental anomaly. Aging has been found to decrease the elasticity of soft tissues causing tendinosis to develop between the stylohyoid ligament and the lesser horn of hyoid bone. This tendinosis causes symptoms mimicking Eagle's syndrome. These patients can hence be labelled as suffering from pseudo stylohyoid syndrome.

Medical management:

Includes local infiltration with hydrocortisone and bupivacaine into the tonsillar fossa transorally. Injection of triamcinolone acetonide (40mg / ml) at the site of maximum tenderness may help in certain patients. Triamcinolone is used for its anti-inflammatory and fibrinolytic effect.

Surgical management: This involves completely breaking and removing a large portion of the elongated stylohyoid component. Two approaches can be used:

1. Intraoral
2. External

Intraoral approach: can be performed after tonsillectomy via the tonsillar bed.

Glogoff procedure:

This is a transpharyngeal procedure to approach styloid process. It is performed under general anesthesia after putting the patient in Rose position (hyperextended and open-mouthed position). If the styloid process could be palpated through the tonsillar fossa, it can be used as a landmark for incising the pharyngeal mucosa. The incision site should be infiltrated with 2% xylocaine mixed with 1 in 100,000 adrenaline in order to reduce mucosal bleeding. A 1 cm long incision is sufficient to expose and remove the styloid process. After slitting the pharyngeal mucosa, the tissue over styloid process is fixed with the help of fingers. Using a Negus knot adjuster, the tissue over the styloid process can be slit open and the periosteum over the styloid process stripped. Once the styloid process has been visualized it can be removed with the help of a rongeur. After securing perfect hemostasis the wound is closed with absorbable sutures.

Another easy intraoral approach is currently being practiced by dental surgeons. In this method the incision is given along the ascending border of the ramus of mandible after infiltrating the area with 2% xylocaine and 1 in 80000 adrenaline. The incision should be deepened by cutting through mucosal and submucosa. By blunt dissection with a curved artery forceps medial to the medial pterygoid muscle and lateral to the superior constrictor muscle the styloid process is exposed. The periosteum is incised without disturbing the attachments to the styloid process and is degloved. The styloid process can easily be removed by fracturing it with an artery forceps.





Image showing the incision for intraoral approach

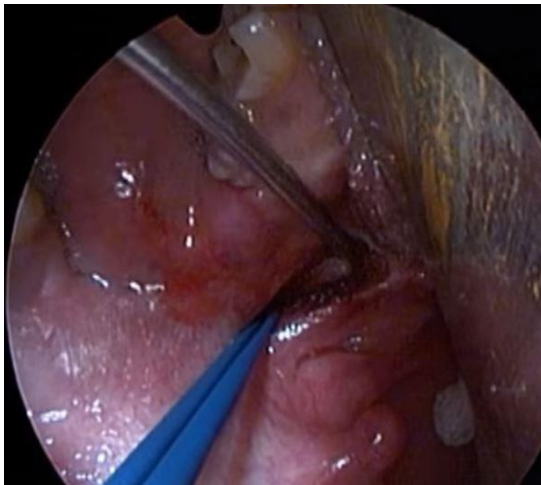


Image showing styloid process exposed

External approach:

This gives excellent exposure and access to the whole of the styloid process. Approach is via a Ridsen incision (submandibular approach). A skin crease incision is made approximately 2 cm below the angle of the mandible. Posterior extension of platysma muscle is identified. Using a combination of blunt and sharp dissection the posterior border of the mandible is exposed. The submandibular gland is dissected and retracted anteriorly. The posterior belly of digastric was dissected and identified, then the same was retracted laterally. The external carotid arterial system is identified and retracted forwards. Digital palpation of the surgical field will reveal the location of the elongated styloid process. The styloid process and stylohyoid ligament were identified after careful dissection. The stylohyoid ligament is

transected at the tip of the styloid process if it is not calcified. If calcified then a bone nibbler needs to be used. The same bone nibbler is used to cut a 2.5 cm segment of the elongated styloid process. Wound is closed in layers. Immediately below the investing fascia under the external carotid or internal maxillary artery the styloid process is identified and exposed. The periosteal lining along with muscle attachments is stripped away from the styloid process. The styloid process is excised and the wound is closed in layers.



Image showing Risdien incision

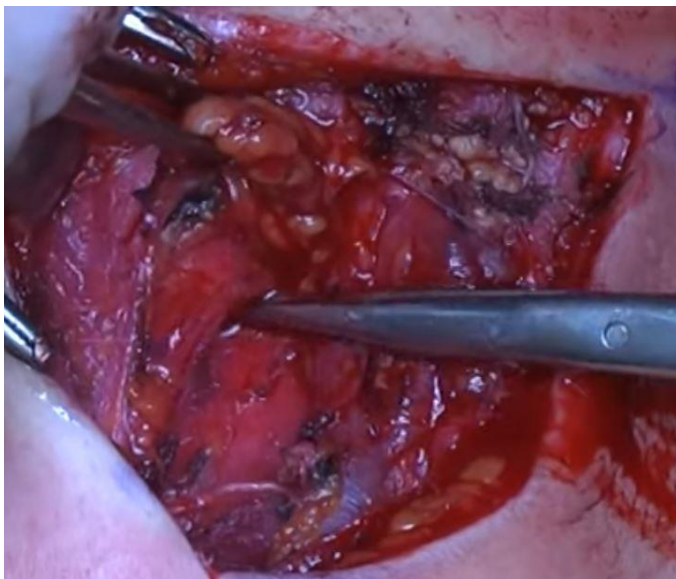


Image showing retraction of submandibular salivary gland

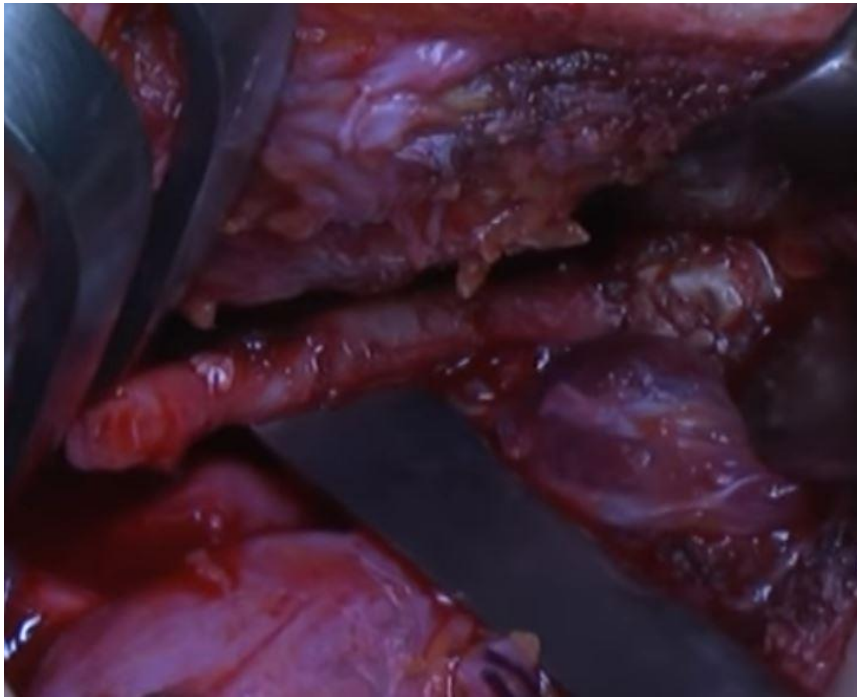


Image showing elongated styloid process exposed

Tonsillar bed approach:

In this approach tonsillectomy is performed first. The muscles of tonsillar bed are dissected and retracted using Negus curved artery forceps. As soon as the styloid process become visible an incision is made at the tip of the styloid process and is stripped using Negus knot adjuster. The elongated styloid process is broken using a bone nibbler and is removed. The wound is closed by interrupted absorbable sutures.