Learning objectives

The purpose of this poster is to:

1) Review the normal appearances of the craniocervical junction and cervical spine (including those anatomical variants also considered 'normal').

2) To illustrate, with the relevant clinical imaging, several cases of torticollis and head-tilt which have been encountered at York District Hospital.
**Background**

Torticollis may be congenital or acquired and is encountered in both paediatric and adult populations. It is characterised by shortening of the sternocleidomastoid muscle and a rotational deformity of the cervical spine which produces a secondary head tilt. Acquired torticollis is often associated with infection or trauma and is more common in older children and adults. In infants and younger children torticollis is more likely to be muscular (e.g. fibromatosis colli) or congenital associated with anatomical anomalies of the craniocervical junction and/or upper cervical spine.

Radiologists play an important role in the diagnosis of cervical spine and neck abnormalities. These pathologies are rare, particularly in a District General Hospital (DGH) setting. When such cases are encountered they can pose significant diagnostic challenge and be a cause of uncertainty unless the radiologist has particular expertise in this area.

This poster has been produced in response to such challenging encounters in patients with both acute and chronic presentation of torticollis and head tilt.

Possible aetiologies include:

<table>
<thead>
<tr>
<th>CONGENITAL</th>
<th>ACQUIRED</th>
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<tbody>
<tr>
<td>Muscular (e.g. hypoplastic SCM)</td>
<td>Trauma</td>
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<td>Bony anomaly (e.g. fusion anomaly)</td>
<td>Atlantoaxial rotatory fixation</td>
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<td>Central Nervous System (e.g. Chiari)</td>
<td>Infection (e.g. parapharyngeal abscess)</td>
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<td>Syndromes (e.g. Down syndrome)</td>
<td>Inflammatory (e.g. JIA)</td>
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<td>Neoplastic (e.g. brain or bone metastases)</td>
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NORMAL APPEARANCES:

In **Fig. 1 on page 6** we demonstrate the 'column' concept of spinal alignment as applicable to the cervical spine. In **Fig. 2 on page 6** we show the normal relationship of the atlantoaxial junction when viewed in the sagittal plane. In **Fig. 3 on page 7** the normal predental space (as measured from the posterior cortex of the anterior arch of C1 to the anterior cortex of the odontoid process of C2). The normal appearances of the C1 and C2 ossification centres are illustrated in **Fig. 4 on page 8**. C1 is formed from three primary ossification centres with fusion of the posterior arch by 5 years and the anterior arch by 8 years. C2 has five primary ossification centres which have usually fused by the age of 12. The craniocervical junction and cervical spine are strengthened by several ligaments, depicted in **Fig. 5**.

PSEUDOSUBLUXATION (**Fig. 6 on page 10**):

2 children with neck pain following minor trauma. Pseudosubluxation is a normal variant seen at C2/C3 or less frequently C3/C4. It is present in up to 46% of children aged <8 years. Spinous process alignment remains normal and the apparent abnormality will 'correct' with extension of the neck.

OS ODONTOIDIUM (**Fig. 7 on page 11**):

16 year old patient. Incidental finding on CT of the orbits. An anatomical variant of C2 believed to be congenital or represent a previously undiagnosed fracture through the odontoid growth plate before age 5 years. May cause instability.

ASYMMETRICAL STERNOCLEIDOMASTOID MUSCLES (**Fig. 8 on page 11**):

A 9 year old with longstanding right sided head tilt. There was a past history of shoulder dystocia at birth which had resulted in a brachial plexus injury. Asymmetry of the sternocleidomastoid muscles was demonstrated on imaging and considered to be causative. He was referred for physiotherapy.

NEONATAL FIBROMATOSIS COLLII (**Fig. 9 on page 12**):

A benign condition which is usually unilateral. There is a slight male predominance. It is due to fibromatosis within the sternocleidomastoid muscle and is believed to be secondary to birth trauma (e.g. forceps delivery). We present selected ultrasound images of two patients with this condition.
KLIPPEL-FEIL SYNDROME (Fig. 10 on page 12):

A 12 year old patient who was referred for ultrasound because of a prominent left sternoclavicular joint. The radiologist identified a scoliosis and arranged radiographs of the spine and thorax. A diagnosis of Klippel-Feil syndrome was ultimately made. This syndrome is due to an error in segmentation which occurs between the 3rd and 8th weeks of gestation. It results in fused cervical vertebrae and is associated with a Sprengel deformity of the shoulder, cervical spondylosis, aortic arch abnormalities and unilateral renal agenesis. The incidence is approximately 1 in 40,000 live births.

ATLANTOAXIAL INSTABILITY (Fig. 11 on page 13):

A 5 year old child with Down Syndrome. Normal anterior distance is 3mm in adults and up to 5 mm in children under 8 years. Ligamentous laxity or bone abnormality leads to excessive movement at the C1/C2 junction. The distance will change on flexion and extension of the neck and neurologic symptoms can occur if there is encroachment on the spinal canal. This condition is also associated with other congenital syndromes including Morquio syndrome, Marfan disease and Neurofibromatosis type 1.

ATLANTOAXIAL ROTATORY SUBLUXATION / FIXATION (AARF):

In AARF the anterior facet of C1 becomes fixed on the ipsilateral facet of C2 and may be associated with dislocation of the lateral mass of C1 on C2.

CASE 1 (Fig. 12): An 8 year old with a 5 month history of painful fixed torticollis. There was a history of torticollis at birth which improved with physiotherapy. In this case the child was referred to the spinal surgeons at Sheffield Childrens Hospital (UK) where the rotational deformity was reduced and internally fixed with an occipital plate and posterior screws from C0 to C2.

CASE 2 (Fig. 13 on page 15): A 2 year old patient currently awaiting spinal surgery.

ANOMALOUS CRANIOCERVICAL JUNCTION:

CASE 1 (Fig. 14 on page 16): Anomalous craniocervical junction in a 20 year old patient.

CASE 2 (Fig. 15 on page 17): Anomalous craniocervical junction and C1 vertebra in a 12 year old patient with longstanding head tilt. The patient has recently been referred to a tertiary paediatric centre for an opinion.
Images for this section:

Fig. 1: sagittal (A) and coronal (B) images. •Anterior column alignment (green line) - smooth arc with no step. •Posterior column alignment (blue line) - smooth arc with no step. •Spinous process alignment (orange line) - smooth arc, no step. •Lateral mass alignment (red lines) - no overlap of C1 on C2.

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Fig. 2: •Occipital condyle (outlined in red) •C1 vertebra (outlined in green) •C2 vertebra (outlines in purple).

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Fig. 3: Normal predental space measuring 3-5mm.

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Fig. 4: Normal ossification centres of C2 in a 4 month old (images A&B). Normal appearances of C1 ossification centres at 1 year (C) and 7 years (D). Normal appearances of the C2 ossification centres at 1 year (E) and 7 years (F)

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Fig. 5: - Anterior atlanto-occipital ligament (blue). - Tectorial membrane (red) - Apical ligament (purple) - Cruciate ligament (green) - Posterior atlanto-occipital ligament (orange)

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Fig. 6: Pseudosubluxation: is a normal variant, most common at C2/3 but also occurs at C3/4. Present in 46% of children under 8 years. Spinous process alignment remains normal and the subluxation will be corrected with extension of the neck.

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Fig. 7: Os Odontoidium: Retrolisthesis of C1 on C2 with angulation of the os so that it abuts the basion of the clivus (A). Os-odontoidium seen in coronal section (B). Incidental additional finding of cervical ribs (C).

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Fig. 8: Asymmetrical Sternocleidomastoid Muscles: Plain radiograph demonstrating head tilt towards right. Selected coronal and transaxial CT images demonstrating a shortened, atrophic right sternocleidomastoid (arrows). Additional incidental finding of an unfused posterior arch of C2 is also noted.

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Fig. 9: Fibromatosis colli. Patient 1 (A&B) showing focal, heterogeneous isoechoic swelling within sternocleidomastoid. Patient 2 (C) demonstrating more fusiform muscle enlargement.

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Fig. 10: Klippel-Feil Syndrome: Radiographs of the Cervical spine (A) and chest (B) demonstrate multiple segmentation defects with hemi-vertebrae, 'butterfly' vertebrae and block vertebral fusion. This is associated with spinal angulation, head-tilt, scoliosis, rib anomalies and a Sprengel deformity of the left shoulder. The MRI (C) demonstrated normal spinal cord and conus and a transitional S1 vertebrae.

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**Fig. 11:** Atlantoaxial Instability: Sagittal T1 MRI (A) narrowing of the spinal canal at C1. The cord is constricted and the odontoid peg hypoplastic. XR Flexion and extension views of the cervical spine (B) demonstrate atlantoaxial instability with anterior displacement of C1. This was treated with surgical fixation (D). Subsequent MRI (C) unfortunately demonstrates that despite fixation, the cord continued to be compromised with development of atrophy and intrinsic high T2 signal.

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**Fig. 12:** Atlantoaxial Rotatory Fixation: Radiograph (A) demonstrating right head tilt. Selected images from the MRI scan (B&C) and multi-planar CT reformats (D) demonstrate rotatory subluxation of C1 on C2. The right lateral mass of C1 (arrow) is dislocated anteriorly relative to the lateral mass of C2 (asterisk). Normal atlanto-occipital joints. Sagittal alignment is preserved on the left. The odontoid peg tilts towards the left with a pressure erosion at the tip.

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Fig. 13: Atlantoaxial Rotatory Fixation: CT with MPR and 3D surface rendering (A) demonstrates the head position to be rotated to the right. (B) There is also left sided C1/C2 malalignment. (Ci) The atlas is aligned with the face but the dens AP axis is rotated anteriorly towards the left. (Cii) The left lateral mass of C1 is subluxed anteriorly relative to the left lateral mass of C2. (Ciii) The axis and rest of the spine are rotated out of alignment to the face.

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Fig. 14: Anomalous Cranio-cervical Junction: CT with MPR - Enlarged left occipital condyle extending postero-inferiorly into the spinal canal (asterisk). The left side of the neural arch is dysplastic, thinned and sclerotic (arrow). The left lateral mass of the atlas is triangular and wedge shaped rather than rhomboid (arrowhead). The elongated left condyle still articulates with the lateral mass of the atlas but extends posteriorly to also articulate with the arch (curved arrow). Pressure erosion of the odontoid peg. Normal C1/C2 joint.

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Fig. 15: Anomalous Cranio-cervical Junction and C1: Abnormal C1 spinous process on lateral radiograph (A). On the CT scan there is an incomplete C1 ring with midline cleft
anteriorly and non-fusion of the posterior arch with the lateral masses (B). The right side of the atlas (B&C) is hypoplastic (asterisk) with asymmetry of the occipital atlantoaxial relationship. The sagittal reformat (D) demonstrates a normal appearance and alignment of the left atlanto-occipital joint. On the right (F) the atlanto-occipital joint is anomalous with the hypoplastic C1 lateral mass extending upwards anterior to the occipital condyle almost to the skull base. There is pressure erosion of the anterior odontoid peg (E) and left occipital condyle (F). No evidence of spinal cord compromise was seen on the MRI scan (G).

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Conclusion

1) Abnormalities of the craniocervical junction and cervical spine can be complex. Radiologists play a crucial role in diagnosing such abnormalities and regardless of speciality should be able to facilitate appropriate imaging in an expeditious manner.

2) Radiologists need to have a sound understanding of the normal appearances of the craniocervical junction and spine, including the 'normal anatomical variants'.

3) Distinguishing clinically urgent and important causes from more indolent and benign causes is important in ensuring patients are managed appropriately.
References

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