Sneeze syncope, basilar invagination and Arnold-Chiari type I malformation

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SYNOPSIS Syncope precipitated by sneezing in an adult male associated with an Arnold-Chiari type I malformation and basilar invagination presents a clinical problem in the differential diagnosis and pathological anatomy of Valsalva-related syncope. An abnormally acute clivoaxial angle, small foramen magnum, and type I Arnold-Chiari malformation appear to be a combination of features intolerant of Valsalva-induced changes in cerebral volume, brain-stem position, CSF fluid dynamics, or blood vessel relationships. Proposed mechanisms of pressure transmission to the area of intracranial pathology are discussed.

CASE REPORT

A 29 year old man (PNH221862) was seen in consultation at the Naval Hospital, Portsmouth, Virginia, with the chief complaint of dizziness associated with sneezes, which had been occurring for the preceding 15 years. Each episode was essentially identical, beginning with a sneeze (although not all sneezes produced the dizziness) and followed immediately by a 'crick' sensation in the back of the neck (the patient could not characterize this sensation more specifically). Severe vertigo then occurred lasting 45 to 60 seconds, followed by nausea which would persist for up to 15 minutes. Nausea,

Sneezing, or any other brief Valsalva manoeuvre may, in rare instances, result in syncope (Engel, 1962). ‘Sneeze syncope’ was the presenting symptom of an adult male who had both basilar invagination and type I Arnold-Chiari malformation. The relationship between these craniovertebral structural anomalies and the physiological events of the Valsalva manoeuvre is the subject of this report.

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FIG 1 Line drawing of the skull and upper cervical spine showing the clivoaxial angle (the angle formed by the plane of the clivus and the axis of the odontoid) to be 120 degrees. Basilar invagination is present with Bull’s angle being 27 degrees and Chamberlain’s line passes through the body of C2 vertebra.
present for only the last three years, was a relatively new feature of the symptom complex. In addition to sneezing, more recently the patient noted that straining during bowel movements and isometric exercises could produce an identical sequence of events. The symptoms were sequentially invariable but the complex occurred unpredictably, in a ratio of about one sneeze (or other Valsalva manoeuvre) in three. Two weeks before his hospitalization while shaving one morning, he sneezed and suffered a typical attack which progressed rapidly from the 'sneeze-and-vertigo phase' to an abrupt, brief loss of consciousness of three to four seconds. For 15 minutes after this episode, any attempt at walking was attended by a tendency to drift to the right 'as though drunk'. Headache, diplopia, dysphagia, hoarseness, weakness of arms or legs were not present then or later. A history was elicited of occasional brief 'electric shock sensations' radiating into both arms on flexion of his head consistent with Lhermitte's phenomenon.

Physical examination revealed a short (170.1 cm), stocky (79.5 kg) man with a thick, short neck. No abnormalities of any of the cranial nerves, strength,
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FIG. 4 The AP laminagram of the craniovertebral junction documents the segmentation error, ‘occipitalization’, of C1 vertebra on the left side (arrow). The odontoid process is seen in the centre (*)

...tone or reflexes, cerebellar or sensory function were found on repeated examinations.

Radiographic examination of the skull and cervical spine revealed partial occipitalization of C1 vertebra, a clivoaxial angle of 120 degrees, and basilar invagination (impression) (Fig. 1). Lamino-...
within the intracranial compartment. In the patient with Arnold-Chiari malformation, this pressure increase presumably results in further herniation of the cerebellar tonsils (Williams, 1972). This phenomenon has been observed during surgery on patients with Arnold-Chiari malformation when cough has produced further herniation of the cerebellar tonsils (Mullan and Raimondi, 1962). Williams's hypothesis, although appealing, was not verified in studies performed by Martins et al. (1972). Patients undergoing myelography were asked to perform a Valsalva manoeuvre when the dye column was at the craniocervical junction and the dye failed to move into the intracranial vault.

Larson et al. (1974) have reported two patients with cough syncope and herniated cerebellar tonsils who were studied with CSF pressure recordings. The authors concluded from limited studies of CSF lumbar pressures that cerebellar herniation was secondary to expansion of cerebral blood volume during cough. Evoked somatosensory potentials were performed before and after coughing. Amplitude suppression during cough and recovery of amplitude after return of CSF pressure to normal suggests that compression of the medulla by herniated tonsils may also contribute to syncope.

The possibility of a strictly mechanical anatomical explanation should also be considered. It has been reported that flexion of the neck in patients with Arnold-Chiari malformation at the time of surgery may result in respiratory arrest (List, 1941; Mullan and Raimondi, 1962). Perhaps other cardiovascular events resulting in syncope could be produced as the result of percussion of the ventral medulla on the clivus or temporary vascular occlusion causing brainstem ischaemia. An abnormally acute clivo-axial angle, small foramen magnum, and type I Arnold-Chiari malformation in concert appear to be anatomical features intolerant of brief Valsalva-induced changes in cerebral volume, brain-stem position, CSF fluid mechanics, or blood vessel relationships.

It is not possible to say with certainty whether mechanical percussion of brain-stem against the anterior lip of the foramen magnum, temporary occlusion of ventral brain-stem vessels, increase in cerebral volume with the Valsalva manoeuvre, or the succession of a CSF pressure wave into a small posterior fossa was responsible for our patient’s symptoms. Any or all of these mechanisms may have been operative.

The opinions expressed are those of the authors and not necessarily those of the Department of the Navy.

REFERENCES