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Incidence of Asymptomatic Chiari Malformation

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Abstract

Aim
The aim of this study is to define the incidence of asymptomatic Chiari malformation in an Irish population.
Methods
MRIs performed over 24 months were analysed. Exclusion criteria include: space occupying lesion, hydrocephalus, Chiari symptoms and inadequate views. Data were analysed to give incidence of asymptomatic Chiari and to analyze the relationship between symptom and position of the cerebellar tonsils (Chi square and Fishers exact test).
Results
Sample Characteristics: 147 patients (Male = 65: Female = 82), age range 15 to 93 years (M_{age} = 53.35, SD= 16.67). 2%had a Chiari malformation (n=2). There was no significant association between symptom and tonsil position (Fishers exact test, χ^2 (8) = 9.98, p = .23.)
Conclusion
This study shows an asymptomatic Chiari Malformation rate of 2%. This study supports the idea that in asymptomatic patients, a tonsil herniation of up to 5 millimeters may be an incidental and inconsequent finding.

Introduction

A Chiari Malformation is defined by herniation of the cerebellar tonsils beyond the foramen magnum of the posterior cranial fossa[1]. This leads to symptoms of headache triggered by actions which raise intracranial pressure such as coughing, sneezing and straining. In 1891 Hans Chiari wrote his first paper on cerebellar tonsil tissue ectopia[2]. He described three classifications. Type 1 was classified as a mild degree of cerebellar tonsil herniation whereby there was no other anatomical abnormality of the cerebellum. It was postulated that although most patients with a type 1 malformation were asymptomatic that in some patients, bulbar compression symptoms could result[2]. The exact definition of a Chiari 1 malformation is debated heavily in the literature, many authors regard a type 1 malformation to be cerebellar herniation of at least 5 millimetres beyond the junction of the foramen magnum[3]. Other authors regard radiological type 1 Chiari malformation to be a herniation of 6 millimetres or more. Unlike Chiari type 2 and type 3, the type one malformations remain asymptomatic until adulthood[4]. Type 2 malformation is defined as displacement of the medulla, fourth ventricle and cerebellar vermis through the foramen magnum. Chiari type 3 malformation is similar to type 2 but with an associated occipital or high cervical encephalocele[4]. Types 2 and 3 are associated with spina bifida, syringomyelia and tend to be symptomatic from birth or early childhood[4].

More recently Chiari 1.5 Malformation has been described which is herniation beyond 12mm[5]. Symptomatic manifestations of tonsillar herniation are usually found with a herniation of 12 mm or more[6]. Symptoms can present in patients with shorter herniation distances which has prompted the demand for research into this area. A 2015 American study by the department of public health in Kent University[7] ran a questionnaire on over seven hundred Chiari type 1 patients. They found that most participants were females aged in their mid-thirties[7]. The most common

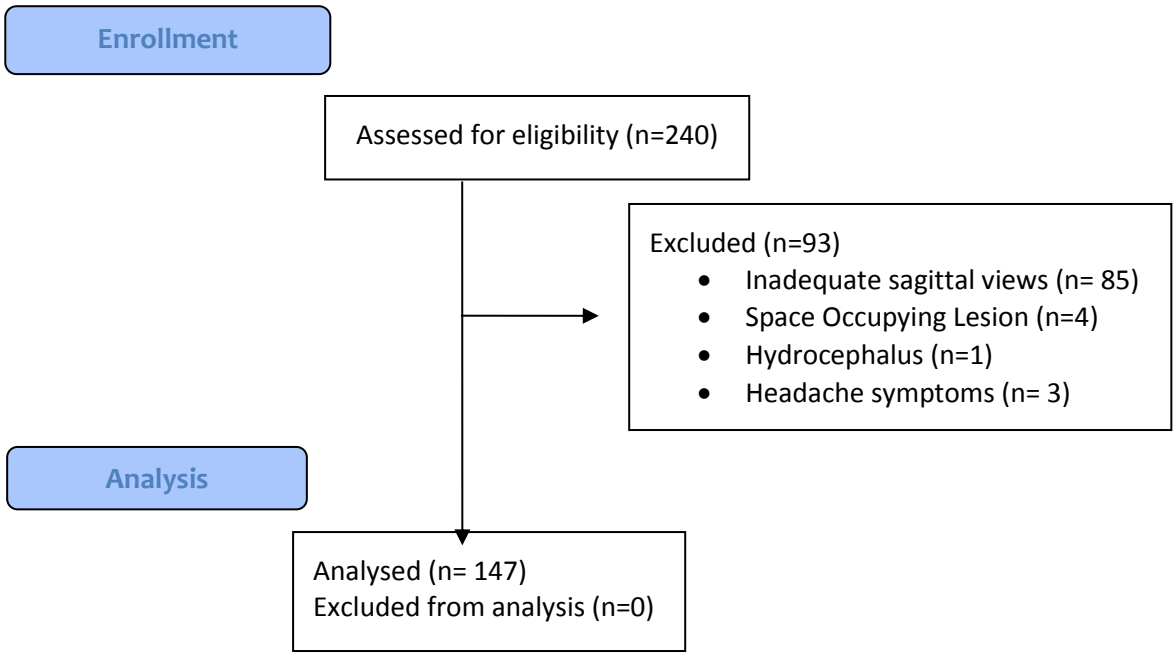
symptom was headache[7]. The average time for a diagnosis was 3.4 years with 24% of those diagnosed being asymptomatic and thus making their diagnosis an incidental finding[7]. The gold standard imaging technique for Chiari Malformation is T1 weighted MRI. American studies have shown the incidence of asymptomatic type 1 Chiari Malformation (>5mm) to be 0.1-1%[8].

The aim of this study is to ascertain the average position of cerebellar tonsils in an asymptomatic Irish population. This may help clarify the prevalence of incidental and asymptomatic cerebellar tonsil herniation in an Irish population.

Methods

This is a retrospective cohort study. Patients whom underwent MRIs to out-rule neurological causes of Hearing Loss (HL), tinnitus, facial palsy or trigeminal neuralgia, were retrospectively selected in a sequential manner over a 24 month period. Indication for scan was based on the electronic referral system, no chart review took place. The MRI protocol was for MRI of the Internal Auditory Meatus (IAM), slice thickness was 1mm. The MRI protocol was for MRI-IAM, slice thickness was 1mm. Select slice, sagittal cut, T1 and T2 weighted MR brain images were analysed for cerebellar tonsil position, a horizontal line was placed at the level of the base of the foramen magnum (from the basion to the opisthion), enabling vertical measurement to the inferior margin of the cerebellar tonsils in millimetres. Exclusion criteria include any scans demonstrating a space occupying lesion, hydrocephalus, Chiari symptoms such as headache and inadequate sagittal views due to artefact.

Fig 1: Participant Flow Diagram



Results

Sample Characteristics
We had a sample of 147 patients (Male = 65: Female = 82), ranging in age from 15 to 93 years (M_{age} = 53.35, SD= 16.67)

MRI Indication
Vertigo was found in 27.9% of the sample (n=41), 25.2% had hearing loss (HL) (n=37), 40.1% had tinnitus (n= 59), 4.1% had facial palsy (n=6), 2.7% had trigeminal neuralgia (n=4).

Cerebellar Tonsil Position
Cerebellar tonsils were found to be at or above the foramen magnum in 71.4% of the sample (n=105). 26.5% of the sample had cerebellar tonsils below the foramen magnum but no chiari (n=39). 2% of the sample had a chiari 1 malformation defined as cerebellar tonsil herniation of 5mm (n=2).

Statistical Analysis

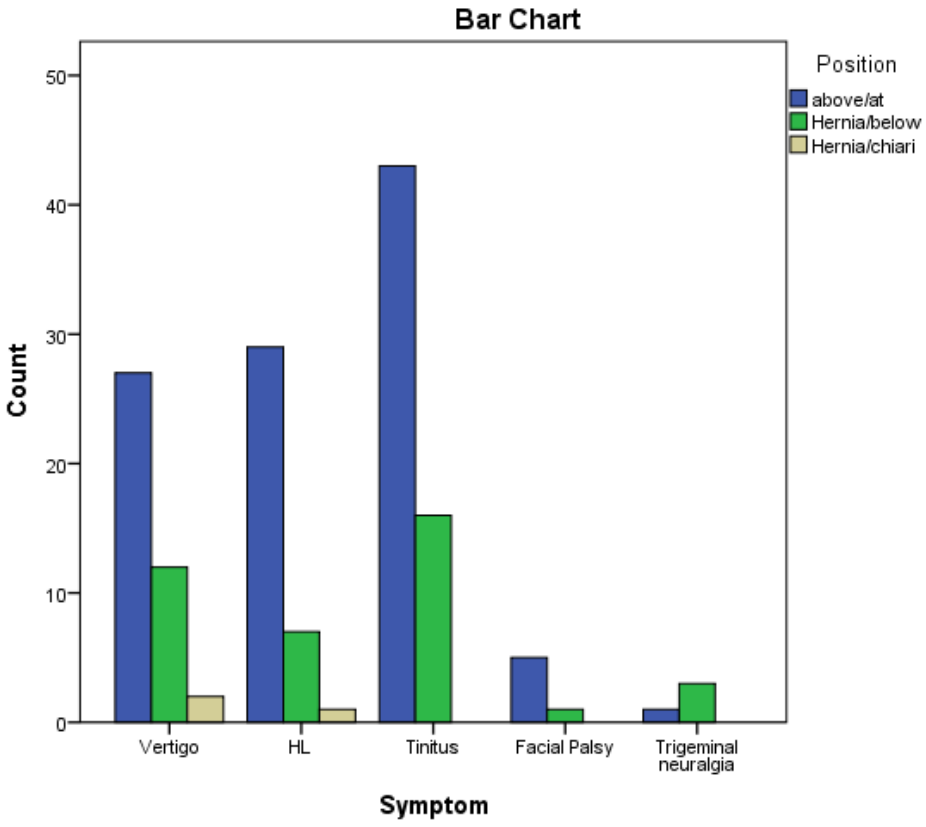
In order to ascertain if there was a relationship between symptom and position of the cerebellar tonsils, we conducted a chi square analysis. The expected frequency in two of the cells was less than 5 and for this reason we report Fishers exact test.

The results of this analysis indicated that there was no association between symptom and position, Fishers exact test, χ^2 (8) = 9.98, p = .23. For each cell the observed frequencies approximated the expected frequencies, with two exceptions; for vertigo, the expected count presenting at Chiari was 8, and the observed count was 2. Similarly, the expected count for vertigo in Chiari was 8 and the observed count was 1 (see Table 1). However, given that fishers exact test was non-significant, we can only assume that this difference does not reflect a difference greater than what could be expected by chance. This being said, given the small numbers in these cells, it's also possible that with a larger sample, these differences may become significant.

Table 1: Expected and observed frequencies in each cell

			Symptom		
			Above/at	Hernia/below	Chiari 1 (5mm herniation)
Symptom	Vertigo	Count	27	12	2
		Expected	29.3	7	8
	HL	Count	29	9.8	1
		Expected	26.4	16	8
	Tinnitus	Count	43	15.7	0
		Expected	42.1	1	1.2
	Facial Palsy	Count	5	1.6	0
		Expected	4.3	3	1
	Trigeminal Neuralgia	Count	1	1.1	0
		Expected	2.9		1

Fig 2: Bar chart of Cerebellar Tonsil position, grouped according to symptoms



Discussion

Asymptomatic Chiari type 1 malformation has not been studied before in an Irish population. An American study performed in John Hopkins in 2000[8] indicates that the incidental rate of asymptomatic Chiari 1 malformation is 0.1-1%[8]. This study had an incidental rate of 2% in a cohort of 147 patients. This study found no statistically significant correlation between scan indication and cerebellar tonsil herniation. The value to this analysis was to evaluate for undiscovered symptoms of Chiari Malformation, away from the typical presentation of headaches. Our study size of 147 is a limiting factor, going forward a larger cohort would strengthen results. In conclusion, the present study shows an asymptomatic Chiari Malformation rate of 2%. We found no correlation between scan indication and tonsil position. In keeping with the clinical context this study supports the idea that in the absence of symptoms, a tonsil herniation of up to 5 millimeters may be an incidental and inconsequent finding. This may help clinicians avoid over investigation of a finding which is not contributing to the clinical picture and will not benefit from aggressive intervention.

Declaration of Conflict of Interest:
The authors declare no conflict of interest.

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