Torticollis in non-syndromal unicoronal craniosynostosis is predominantly ocular-related

Masterthesis, Clinical Sciences for Health Professionals, Program in Clinical Health Sciences, University Medical Center Utrecht, Utrecht University, The Netherlands

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Abstract

Background: Patients with unicoronal craniosynostosis (UCS) often show torticollis, which can result from either an ocular cause or an imbalance in the neck muscles. For the primary caregiver, the plastic surgeon, it is crucial to know the prevalence of ocular torticollis (OT) to ensure appropriate referrals for treating torticollis to an orthoptist or physiotherapist. Furthermore, associated ophthalmic features with OT in these patients are scarcely described. Understanding these associations helps orthoptists in developing effective treatment plans for these patients.

Purpose: To determine the prevalence of OT in a population-based cohort of UCS patients and investigate its association with binocular single vision (BSV), strabismus, ocular motility, cyclotorsion, refractive error, and amblyopia.

Methods: In this descriptive cross-sectional study, medical records of non-syndromic UCS patients treated between 1994-2022 at a tertiary-care hospital, were retrospectively reviewed. Collected data included diagnosis and type of torticollis, BSV, strabismus, ocular motility, alphabetical patterns, cyclotorsion, refractive error, and amblyopia. Patients were classified as OT based on their ophthalmic and/or orthoptic diagnosis. Prevalence was determined using a binominal test and its confidence interval using the Clopper-Pearson test. Associations between OT and the ophthalmic features were determined using Chi-square or Fishers exact test and the effect size was calculated using Cramer's V.

Results: In total 146 patients with UCS were included; of whom 57 had torticollis. An ocular cause for the torticollis was found in 54 patients 37% (95% CI [0.292 – 0.454]). Significant associations were found between OT and strabismus ($p<0.001$), ocular motility abnormalities ($p<0.001$), alphabetical patterns ($p<0.001$), and amblyopia ($p=0.002$). BSV ($p=0.277$) and refractive error ($p=1.0$) were not significantly associated with OT. Although, in OT the BSV was relatively poor (42.1%) and more frequently absent (26.3%) compared to the non-torticollis group (7% and 16.3%, respectively). In both groups excyclotorsion was predominantly present (62.3%).

Conclusion: In 95% of cases, torticollis in UCS patients is ocular-related. Overall, one-in-three patients with UCS have OT.

Recommendations: This study emphasizes the importance of plastic surgeons referring all UCS patients with torticollis to an orthoptist, who specializes in diagnosing and treating OT, before considering physiotherapy.

Keywords (5)
Unicoronal craniosynostosis, torticollis, prevalence, ocular features, plagiocephaly

Nederlandse samenvatting

Achtergrond: Patiënten met unicoronale craniosynostose (UCS) vertonen vaak torticollis, die kan voorkomen door een oculaire oorzaak of een onbalans in de nekspieren. Het kennen van de prevalentie van een oculaire torticollis (OT) is van belang voor plastisch chirurgen - als hoofdbehandelaar - om een passende doorverwijzing voor de behandeling van torticollis te faciliteren; hetzij naar een orthoptist of fysiotherapeut. Geassocieerde oogheelkundige kernmerken van OT zijn beperkt beschreven. Het kennen van deze associaties helpt orthoptisten bij het ontwikkelen van effectieve behandelplannen voor deze patiënten.

Doel: Het bepalen van de prevalentie van OT in een populatie gebaseerd cohort van UCS-patiënten en bepalen van de associatie met binoculair enkel zien (BEZ), scheelzien, oogmotiliteit, cyclotorsie, refractie fout en amblyopie.

Methode: In deze beschrijvende cross-sectionele studie werden medische dossiers van patiënten met non-syndromale UCS, behandeld in een ziekenhuis voor tertiaire zorg, retrospectief beoordeeld. Verzamelde gegevens omvatten de diagnose en het type torticollis, BEZ, scheelzien, oculaire beweeglijkheid, alfabetische patronen, cyclotorsie, refractieve afwijking en amblyopie. Patiënten werden geclassificeerd als OT gebaseerd op de oogheelkundige en/of orthoptische diagnose. De prevalentie werd bepaald met een binominale test en zijn betrouwbaarheidsinterval met de Clopper-Pearson test. Associaties tussen OT en oogheelkundige kenmerken werden bepaald met de Chi-kwadraattoets of Fishers exact toets, effectgroottes werden berekend met Cramer's V.

Resultaten: Er zijn 146 UCS-patiënten geïncludeerd; 57 hadden een torticollis. Een oculaire oorzaak voor de torticollis werd gevonden bij 54 patiënten 37% (95% CI [0.292 – 0.454]). Significante associaties werden gevonden tussen OT en scheelzien ($p<0.001$), afwijkingen in oogmotiliteit ($p<0.001$), alfabetische patronen ($p<0,001$) en amblyopie ($p=0.002$). BEZ ($p=0.277$) en refractieve afwijkingen ($p=1.0$) waren niet significant geassocieerd met OT. Hoewel het BEZ in OT relatief slechter was (42.1%) en vaker afwezig (26.3%) in vergelijking met de groep zonder torticollis (respectievelijk 7% en 16.3%). In beide groepen was een excyclotorsie overwegend aanwezig (62.3%).

Conclusie: In 95% van de gevallen heeft torticollis bij UCS-patiënten een oogheelkundige oorzaak**.** In totaal heeft één-op-de-drie UCS-patiënten OT.

Aanbevelingen: Dit onderzoek benadrukt het belang van plastisch chirurgen om alle UCS-patiënten met torticollis te verwijzen naar een orthoptist, die gespecialiseerd is in het diagnosticeren en behandelen van OT, alvorens een verwijzing naar fysiotherapie.

Introduction

Craniosynostosis is a rare congenital condition characterized by the premature closure of one or more cranial sutures[1]. It can occur as part of a syndrome or as an isolated finding (non-syndromic)[2,3]. Unicoronal craniosynostosis (UCS) is a non-syndromic form, in which one coronal suture closes prematurely[3,4]. It is also known as anterior plagiocephaly and accounts for 30-35% of non-syndromic craniosynostosis cases[5]. The prevalence of UCS is approximately 1 in 10,000 live births worldwide, with a higher incidence in females[4,6]. Due to the premature closure of the coronal sutures, the infant's head growth is restricted at the side of the closure, leading to growth occurring on the ipsilateral side. This results in several skull deformations: a flattened forehead and base of the orbit on the side of the fusion (retrusion of the forehead and superior orbital rim), protrusion of the forehead on the contralateral side (frontal bossing of the forehead), raised eye socket on the ipsilateral side (orbital asymmetry), and a shift of the nose towards the ipsilateral side (nasal deformity)[7]; *figure 1*. Surgical correction by a plastic surgeon is typically required to address the suture closure. Ocular abnormalities associated with UCS include strabismus (misalignment of the eyes), aniso-astigmatism (different amounts of astigmatism between both eyes), and amblyopia (lazy eye)[8]. Torticollis, characterized by head tilt, can also occur in UCS, either as ocular-related or non-ocular-related torticollis[9–12].

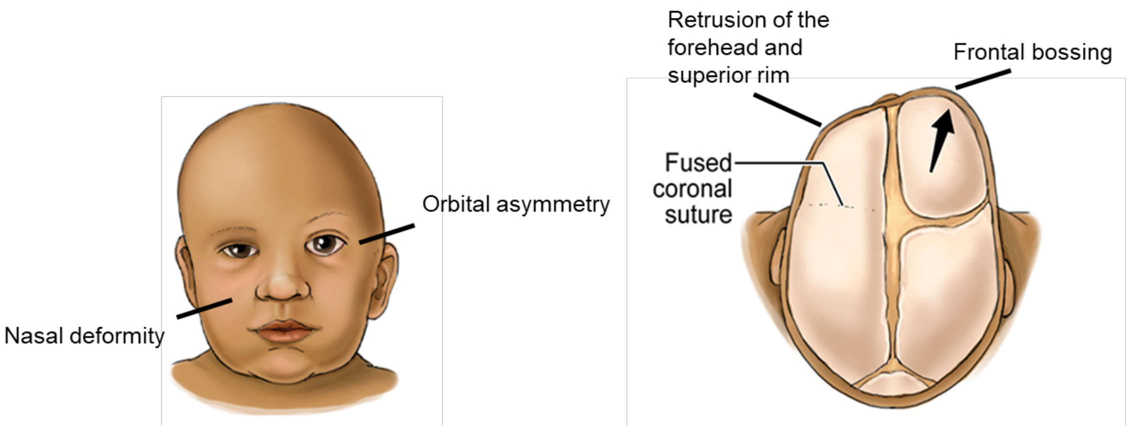

Figure 1: Unicoronal suture synostosis cause a suture-specific skull shape and deformations. (Adjusted from Mount Sinai Craniofacial Clinic[13], 2023)

The prevalence of ocular torticollis (OT) in patients with UCS is unknown, despite being a commonly reported feature[11,14]. Determining this prevalence is crucial for plastic surgeons, as primary caregiver, to make appropriate referrals for treating torticollis. Non-ocular torticollis in UCS is commonly caused by sustained contraction of the sternocleidomastoid muscle[15]. In these cases, physiotherapy is needed to prevent permanent torticollis by relieving tense neck muscles and achieving an upright head position[9,10,12,16]. On the contrary, OT is an abnormal head posture resulting from an ocular cause. It serves as a compensatory mechanism to alleviate double vision caused by strabismus and obtain binocular single vision (depth perception) (BSV) or to improve visual acuity (eyesight)[17]. Maintaining OT is vital for preserving BSV[17–20]. If no OT is not maintained, it can lead to double vision, suppression of the squinting eye, and the development of amblyopia[20,21]. Referral to an orthoptist is essential for treating OT[18], as their expertise is to effectively resolve OT through strabismus surgery[18,21].

The prevalence of strabismus in UCS patients, especially incomitant strabismus, is significantly higher (40-70%) [22–28] compared to the general population (1.93%)[29]. Incomitant strabismus is a type of strabismus where the misalignment of the eyes varies with the direction of gaze[30]. This high prevalence is hypothesized as the ocular reason why these patients adopt torticollis[11,31]. Strabismus in UCS can result from the abnormal position of the trochlea or the absence of the superior oblique muscle[25,28] *(Figure 2A).* This can lead to excyclotorsion syndrome, also known as pseudo-superior oblique palsy or strabismus sursoadductorius[32,33]. The excyclotorsion syndrome causes several ocular motility abnormalities to occur: elevation in adduction, V-pattern strabismus, and excyclotorsion[11,31] *(Figure 2B-D).* Unfortunately, little is known about these associated ophthalmic features in UCS patients with OT compared to UCS patients without OT[11].

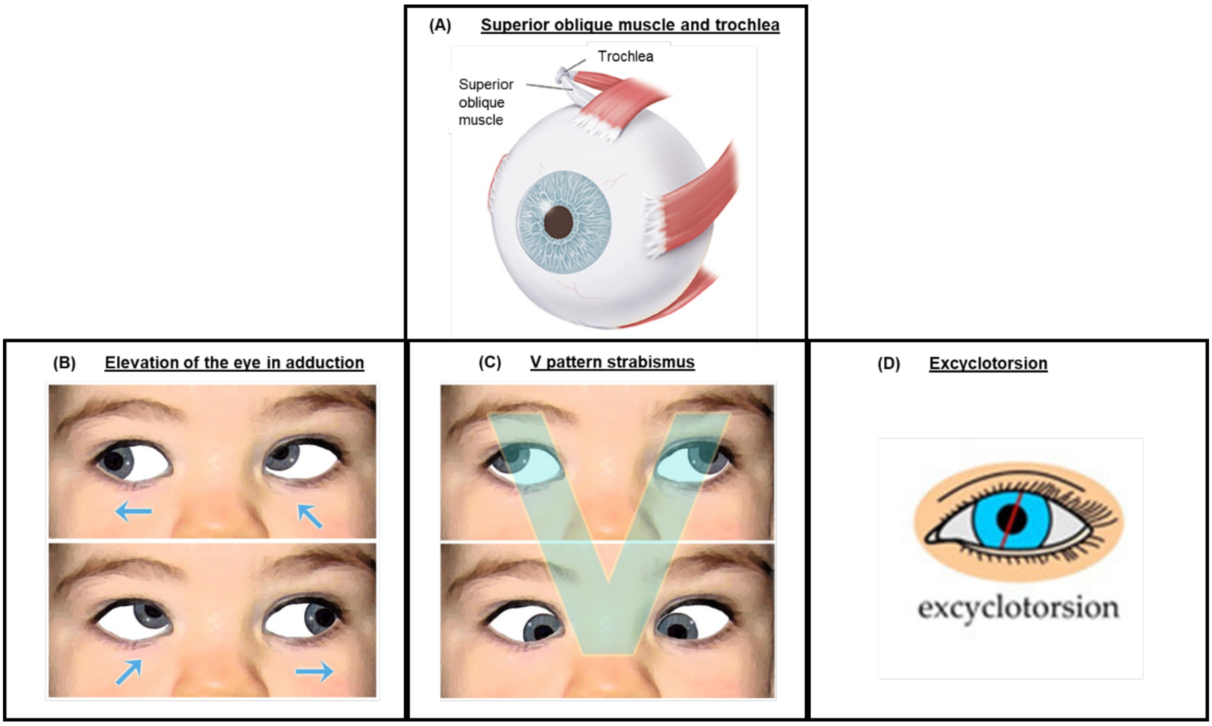

Figure 2: (A) Showing the superior oblique muscle and trochlea. If the position of the trochlea is abnormal or if the superior oblique muscle is absent this leads to an excyclotorsion syndrome.Excyclotorsion syndrome causes (B) elevation in adduction (elevation of the eye when looking towards the nose), (C) V pattern strabismus (outward squint when looking upwards, together with an inward squint when looking downwards) and (D) excyclotorsion (outward rotation of the eye). (Adjusted from Saint Luke's[34] 2023, Athens eye hospital[35,36] 2023, Pencic et al.[37] 2022)

Determining the prevalence of OT and its associated ophthalmic features in a large population-based cohort of UCS patients is vital. This information is necessary for plastic surgeons to make appropriate referrals for treating torticollis in UCS patients, whether to an orthoptist or physiotherapist. For orthoptists, knowing its associated ophthalmic features allows the development of effective treatment plans for these patients.

Aim

The primary aim of this study is to determine the prevalence of OT in a large population-based cohort of patients diagnosed with UCS. Second, this study aims to investigate the association between the presence of OT and BSV, strabismus, ocular motility, the presence of cyclotorsion, amblyopia, and refractive error among these patients.

Methods

Study design

In this retrospective descriptive population-based cross-sectional study, the medical records of patients with UCS treated between 1994 and 2022 at a tertiary-care hospital (Erasmus MC, Rotterdam, the Netherlands) were reviewed. This is the designated center in the Netherlands for craniofacial surgery, so all patients with craniofacial disorders are referred here. Therefore, the sample of patients is representative of the population of interest. Clinical data from 1994 onwards were electronically available and thus traceable. Children diagnosed with UCS typically undergo surgery to correct for the suture closure between 6-9 months of age. Afterward, ophthalmologic screening for the presence of papilledema is a part of the multidisciplinary craniofacial assessment in these patients until 18 years of age (*Figure 3*). The current treatment guideline[38] used in the Netherlands indicates referral to the orthoptist if considered necessary by the plastic surgeon.

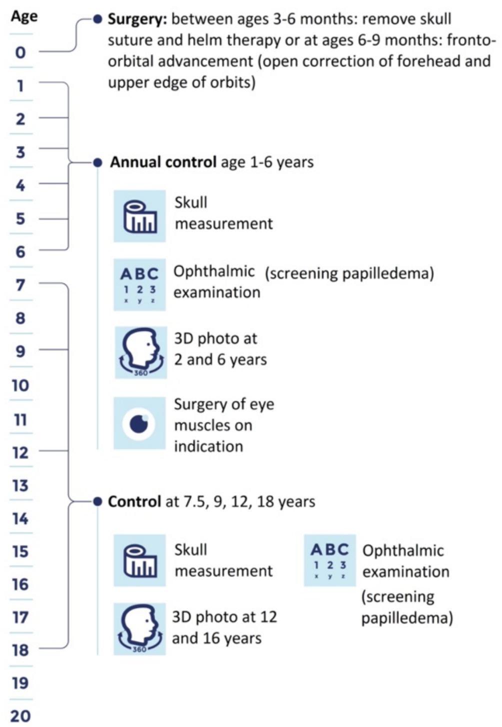

Figure 3 Flowchart of follow-up routine in UCS patients at the Erasmus MC, Rotterdam, the Netherlands

Participants

With the help of the Department of Research Suite at the Erasmus MC, patients diagnosed with UCS between 1994 and 2022 were identified. Patients diagnosed with non-syndromal UCS, confirmed through CT-scans assessed by a radiologist, medical examinations, and confirmed by genetic testing were considered eligible. Patients were included if surgical, ophthalmologic, or orthoptic assessments were available. Patients with a history of strabismus surgery elsewhere were excluded from the study, as this could potentially affect or resolve an observed torticollis in these patients. Patients were categorized into two groups to compare their ophthalmic features in relation to OT: UCS patients with OT and those without. Patients with non-ocular torticollis were included in the group without torticollis.

Variables

To avoid observer bias, the researcher (ET) followed a standardized protocol while extracting data from medical records. The collected information for each patient included their sex, side of synostosis, type, age at craniofacial surgery, and age at the initial visit. Clinical data from the initial ophthalmologic and/or orthoptic visit were collected, which included the following: 1) presence or absence of observed torticollis and its type, 2) BSV, 3) presence of strabismus, 4) ocular motility, 5) presence or absence of cyclotorsion, 6) refractive error, and 7) presence or history of amblyopia. All clinical data, except for cyclotorsion, were obtained from the initial visit. If cyclotorsion was not determined during the initial visit, the first determination prior to strabismus surgery was considered. Incomplete examinations were included as missing data.

Orthoptic measurements

The variables were determined based on the orthoptic measurements as described below:

1) The presence of torticollis was collected by reviewing the records of the orthoptist and ophthalmologist for the description of observed torticollis (yes/no). Also, the records of the Department of Plastic Surgery were scanned for descriptions of torticollis. The type of torticollis was defined as a face turn, chin elevation, chin depression, head-tilt or, a combination of the aforementioned (*Figure 4*). Patients were considered to have OT based on their ophthalmic and/or orthoptic diagnosis.

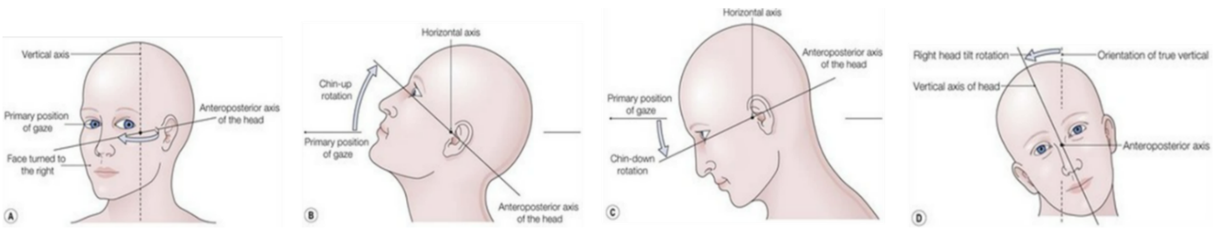

Figure 4 Shows the types of torticollis. (a) Face turn to the right: the head is rotated to the right side of the primary gaze in the vertical axis. (b) Chin elevation: the chin is elevated relative to the primary position in the horizontal axis. (c) Chin depression: the chin is depressed relative to the primary position in the horizontal axis. (d) Head-tilt to the right shoulder: the head is tilted to the right shoulder in the anteroposterior axis (Adjusted from Kraft[39], 2016).

2) The BSV was determined by measuring stereopsis, which is a sensitive measure of binocular vision[40–42]. Stereopsis was assessed the following stereotests: Bagolini Straited Glasses (positive or negative outcome). Or the Lang, Titmus housefly, and/or TNO tests (outcomes measured in arcseconds). Outcomes were categorized as 'not present' if Bagolini was negative, 'poor' if Bagolini was positive and Titmus housefly

measured 3.772 arcseconds, 'moderate' if Titmus circles 200-40 arcseconds were recognized, and 'good' if all figures of the Lang Test (200 arcseconds) or TNO plate V (480-240 arcseconds), VI or VII (120-15 arcseconds) were positive[43].

3) The angle of strabismus (horizontal- and vertical or combined deviations) was measured using the prism cover test at near (30cm) in prism degrees. Outcomes were categorized as esotropia, exotropia, esotropia and vertical deviation, exotropia and vertical deviation, vertical deviation alone, or not present.
4) Ocular motility was assessed by observing the eye movements while the patient followed a moving penlight from the primary position to each gaze position[30]. Motility abnormalities were described as elevation or depression in adduction of one or both eyes, if present. The presence of an alphabetical pattern, defined as a V- or A-strabismus pattern or not present, was also noted during the assessment of ocular motility.
5) Cyclotorsion was measured using the Maddox Double Rod test or assessed anatomically by the ophthalmologist during fundus examination. Outcomes were defined as incyclotorsion, excyclotorsion, or not present.
6) Refractive errors were obtained by retinoscopy in cycloplegia. Outcomes were defined as hypermetropia (≥+1.0D), high hypermetropia (≥+5.0D), myopia (≥-1.0D), high myopia (≥-5.0D), astigmatism (≥-1.0D), anisometropia (≥1.0D).
7) The presence of amblyopia was defined as visual acuity (VA) difference of ≥0.2LogMAR between both eyes and was categorized as present or not present. VA was measured using the Lea Hyvarine Chart or numbers/letters on the projector, expressed in LogMAR.

Study size

The Department of Research Suite identified a total of 277 patients diagnosed with UCS. Based on this population size, a sample size of ≥159 patients are needed to determine the prevalence of torticollis with a confidence level of 95% with a margin of error of 5 with a population proportion of 50%. Since the prevalence of OT in UCS patients is unknown, it is recommended in the literature to assume a 50% proportion when calculating the required sample size[44,45].

Statistical methods

Descriptive statistics were analyzed using SPSS V.26 (IBM Corps). The prevalence of torticollis in patients with UCS was computed using a binominal test dividing the total number of patients that had OT by the total number of patients in the cohort. The 95% CI was determined using Clopper-Pearson exact test. To characterize the ophthalmic features of UCS patients with and without OT, descriptive statistics were conducted. Age was considered a quantitative continuous variable; results were presented as means with a standard deviation. The independent samples *T*-test was used to compare means between groups. Levene's Test was used to test for homogeneity of variance. The results of each qualitative variable were presented by frequency (n, %). All ophthalmic features were categorical variables, so associations between OT and the features were investigated using either Chi-square or Fisher's exact test. If the expected frequency in each cell of the contingency table was ≥5, a chi-square test was used to verify the association. If the expected frequency in each cell was <5 in >20% of cells, a Fisher's exact test was used. Also, Cramer's V test was performed to calculate the effect size for both the chi-square and Fisher's exact test: a value closer to 1 indicates a greater effect size. Depending on the degrees of freedom, the effect size was rated as small, moderate, or strong[46]. Significant associations underwent further analysis to identify key contributing features to torticollis using a logistic regression model for variables with a binary outcome. In all analyses, $p<0.05$ was considered statistically significant.

Reporting and ethical issues

The Strengthening the Reporting of Observational Studies in Epidemiology Statement (STROBE) for cross-sectional studies was used as transparency reporting guideline. The study was conducted in accordance with the principles of the Declaration of Helsinki (2013). Since it was part of a larger observational longitudinal study approved by the Erasmus MC's ethical board (MEC-2022-0367), this study was not subject to the Medical Research Involving Human Subjects Act (WMO).

Results

Participants

A total of 277 children diagnosed with UCS were treated at the Erasmus MC; 146 met the inclusion criteria. Most patients (n=114) were excluded due to missing orthoptic assessments, due to multi-suture or other craniofacial disorders (n=13), due to an unconfirmed diagnosis of UCS (n=2), due to the presence of Graves' Orbitopathy which could influence the ocular motility (n=1) or due to a history of strabismus surgery (n=1) *(Figure 5).*

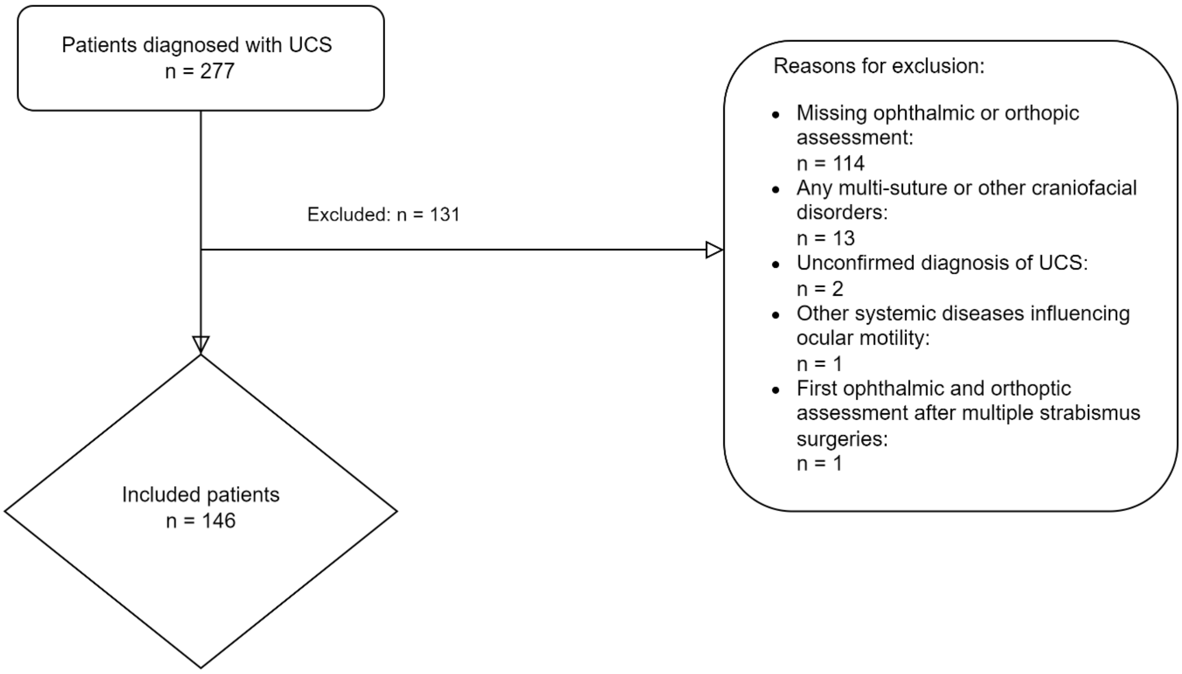

Figure 5: Flowchart of included patients and reasons for exclusion

Of the included patients 96 (65.8%) were female. The right coronal suture was closed in 85 (58.2%) patients. The mean age at the initial visit was 2.72 years (SD 3.6, range 0-27 years). No significant differences were observed in patients' characteristics between patients with and without torticollis (*Table 1)*.

Table 1: Patient characteristics of included UCS patients (n=146)

Patient characteristics	**Total n=146**	**Torticollis n=57**	**No torticollis n=89**	**p-value**
Sex (n, %)				0.853
Female	96 (65.8)	38 (66.7)	58 (65.2)	
Male	50 (44.2)	19 (33.3)	31 (34.8)	
Synostotic coronal suture (n, %)				0.428
Right	85 (58.2)	31 (54.4)	54 (60.7)	
Left	58 (39.7)	25 (43.9)	33 (37.1)	
Unknown	3 (2.1)	1 (1.8)	2 (2.2)	
Age at initial visit (years, mean (SD))	2.72 (SD 3.6)	2.3 (SD 2.5)	3.0 (SD 4.1)	0.256 [95%CI -0.51; 1.91]
Craniofacial surgery (n, %)				0.201
Fronto-orbital advancement rim	135 (92.5)	52 (91.3)	83 (93.3)	
Right and left orbit	*5 (3.7)*	*2 (3.8)*	*3 (3.6)*	
Right orbit	*77 (57)*	*28 (53.8)*	*49 (59)*	
Left orbit	*50 (37)*	*21 (40.5)*	*29 (34.9)*	
Unknown	*3 (2.3)*	*1 (1.9)*	*2 (2.5)*	
Strip craniotomy	1 (0.7)	0	1 (1.1)	
No surgery	5 (3.4)	4 (7)	1 (1.1)	
Unknown	5 (3.4)	1 (1.7)	4 (4.5)	
Mean age at craniofacial surgery (months, mean (SD))	11.58 (SD 7.3)	10.49 (SD 3.0)	12.26 (SD 8.8)	0.087 [95%CI -0.78; 4.32]

Main results

Primary outcome: the prevalence of torticollis

Of the 146 patients identified with UCS, 57 patients had torticollis. Of the 57 patients identified with torticollis, 94.7% (n=54) were ocular-related. The prevalence of OT in patients with UCS was found to be 37% (95% CI [0.292 – 0.454]). Mean age of consultation at which torticollis was identified was 2.8 years (SD 2.6, range 9 months-12 years).

The most common diagnosis for OT was strabismus (93%, n=53), of which 88.7% were incomitant (*Table 2)*. The majority showed a pseudo-superior oblique palsy (59.6%), followed by pseudo-inferior oblique palsy (12.3%), accommodative esotropia (5.3%), and infantile esotropia combined with pseudo-superior oblique palsy (5.3%). The type of torticollis was specified in 56 patients: a head tilt was present in 47.4%, a face turn in 28.1%, or a combination in 22.8% (*Table 3)*. Torticollis with only a chin elevation or depression was not present.

Table 2: Diagnosis in patients with torticollis (n=57)

Diagnosis of torticollis	***n = 57* *[n, %]***
Incomitant strabismus	**47 (88.7)**
Pseudo-superior oblique palsy	34 (59.6)
Pseudo-inferior oblique palsy	7 (12.3)
Infantile esotropia and pseudo-superior oblique palsy	3 (5.3)
Fourth nerve palsy	1 (1.8)
Intermittent exotropia and pseudo-inferior oblique palsy	1 (1.8)
Infantile exotropia and pseudo-superior oblique palsy	1 (1.8)
Concomitant strabismus	**6 (10.5)**
Accommodative esotropia	3 (5.3)
Intermittent exotropia	2 (3.5)
Micro esotropia	1 (1.8)
Other	**1 (1.7)**
Congenital nystagmus	1 (1.8)
Non-ocular	**3 (5.3)**

Table 3: Described type of torticollis (n=57)

Type torticollis	***n = 57* *[n, %]***
Face turn	**16 (28.1)**
Right	4 (7.1%)
Left	12 (21.4%)
Head tilt	**27 (47.4)**
Right	16 (28.6%)
Left	11 (19.6%)
Combination	**13 (22.8)**
Turn, chin, tilt	4 (7.1%)
Turn, chin	3 (5.4%)
Chin, tilt	2 (3.6%)
Tilt, turn	4 (7.1%)
Unspecified	**1 (1.7)**

Secondary outcomes: The ophthalmic-related features associated with OT

Collected ophthalmic-related features and their frequencies are shown in *Table 4*, which features are described in detail below. *Table 5* compares the occurrence of these features in UCS patients with or without OT, their association with the presence of OT, and the effect size (V).

Table 4: Distribution of the ophthalmic related features in the total group, patients with ocular torticollis and patients without (ocular) torticollis.

Ophthalmic features	*Total* [n, %]	*Ocular torticollis* [n, %]	*No (ocular) torticollis* [n, %]
Binocular single vision	***n = 62***	***n = 19***	**n = 43**
Good	29 (46.8)	3 (15.8)	26 (60.5)
Moderate	10 (16.1)	3 (15.8)	7 (16.3)
Poor	11 (17.7)	8 (42.1)	3 (7)
Not present	12 (19.4)	5 (26.3)	7 (16.3)
Strabismus	***n = 145***	***n = 53***	***n = 92***
Esotropia	11 (7.6)	4 (7.5)	7 (7.6)
Exotropia	8 (5.5)	2 (3.8)	6 (6.5)
ET + vertical	19 (13.1)	10 (18.9)	9 (9.8)
XT + vertical	14 (9.7)	11 (20.8)	3 (3.3)
Vertical alone	17 (11.7)	13 (24.5)	4 (4.3)
Not present	76 (53.8)	13 (24.5)	63 (68.5)
Ocular motility	***n = 143***	***n = 53***	***n = 90***
Elevation in adduction RE	26 (18.2)	15 (28.3)	11 (12.2)
Elevation in adduction LE	20 (14)	12 (22.6)	8 (8.9)
Elevation in adduction RLE	23 (16.1)	14 (26.4)	9 (10)
Depression in adduction RE	6 (4.2)	5 (9.4)	1 (11.1)
Depression in adduction LE	5 (3.5)	2 (3.8)	3 (3.3)
Depression in adduction RLE	2 (1.4)	2 (3.8)	0
Not present	61 (42.7)	3 (5.7)	58 (64.4)
Alphabetical pattern	***n = 143***	***n = 53***	***n = 90***
V-pattern	46 (32.2)	29 (54.7)	17 (18.9)
A-pattern	12 (8.4)	7 (13.2)	5 (5.6)
Not present	85 (59.4)	17 (32.1)	67 (75.6)
Cyclotorsion	***n = 53***	***n = 30***	***n = 23***
Excyclotorsion	33 (62.3)	22 (73.3)	11 (47.8)
Incyclotorsion	4 (7.5)	1 (3.3)	3 (13)
Not present	16 (30.2)	7 (23.3)	9 (39.1)
Refractive error*	***n = 125***	***n = 45***	***n = 80***
Hypermetropia	112 (89.6)	41 (89.1)	71 (89.9)
High hypermetropia	6 (4.8)	2 (4.4)	4 (5)
Myopia	2 (1.6)	1 (2.2)	1 (1.3)
High myopia	0	0	0
Astigmatism	55 (44)	20 (43.5)	35 (44.3)
Anisometropia	33 (22.6)	12 (26.7)	21 (26.3)
Amblyopia	**n = 146**	**n = 54**	**n = 92**
Present	64 (43.8)	33 (61.1)	31 (33.7)
Absent	82 (56.2)	21 (38.9)	61 (66.3)

Abbreviations: ET+vertical: esotropia combined with a vertical deviation, XT+vertical: exotropia combined with a vertical deviation, RE: right eye, LE: left eye
*The cumulative percentage exceeds 100% since refractive error can occur combined: spherical (hypermetropia/myopia) and/or astigmatism and/or the presence of an anisometropia

Table 5: Associations between the presence of the investigated ophthalmic features and the presence of a torticollis in UCS patients.

Ophthalmic features	***Ocular torticollis*** **[n, %]**	***No (ocular) torticollis*** **[n, %]**	**p-value**	**Cramers' V**
Binocular single vision	***n = 19***	***n = 43***	*0.277*	*0.117*
Present	*14 (73.7)*	*36 (83.7)*		
Absent	*5 (26.3)*	*7 (16.3)*		
Strabismus	***n = 53***	***n = 92***	*<0.001*	*0.406*
Present	39 (73.6)	29 (31.2)		
Absent	14 (26.4)	63 (68.8)		
Ocular motility abnormalities	***n = 53***	***n = 90***	*<0.001*	*0.574*
Present	50 (94.3)	32 (35.6)		
Absent	3 (5.7)	58 (64.4)		
Alphabetical pattern	***n = 53***	***n = 90***	*<0.001*	*0.428*
Present	36 (67.3)	22 (24.4)		
Absent	17 (32.7)	68 (75.6)		
Refractive error*	***n = 45***	***n = 80***		
Hypermetropia	43 (95.6)	75 (93.8)	1.0	-
Myopia	1 (2.2)	1 (1.2)	1.0	-
Astigmatism	20 (44.4)	35 (38.9)	1.0	-
Anisometropia	12 (26.7)	21 (26.2)	1.0	-
Amblyopia	***n = 54***	***n = 92***	*0.002*	*0.267*
Present	33 (61.1)	31 (33.7)		
Absent	21 (38.9)	61 (66.3)		

* The cumulative percentage exceeds 100% since refractive error can occur combined: spherical (hypermetropia/myopia) and/or astigmatism and/or the presence of an anisometropia. In this analysis 'hypermetropia' is cumulative of both the patients with 'hypermetropia' and 'high hypermetropia', and 'myopia' is cumulative of both the patients with 'myopia' and 'high myopia'.

Binocular single vision

BSV was determined in 19 out of 54 patients with OT and in 43 out of 92 patients without (ocular) torticollis. In the OT-group, BSV was present in 73.7% of patients compared to 83.7% of patients in the non-torticollis (NT) group. There was no significant association between BSV and OT presence (p=0.277) (*Table 5)*. In the OT-group, BSV was relatively poor (n=8, 42.1%) and more frequently absent (n=5, 26.3%) compared to the NT-group (n=3, 7% and n=7, 16.3%, respectively) *(Table 4).*

Strabismus

In 53 patients with OT and 92 patients without (ocular) torticollis, the presence of strabismus was assessed. In the OT-group, strabismus was observed in 73.6% of patients, compared to 31.2% in the NT-group. The association between strabismus and OT was moderate (V=0.406) and statistically significant ($\chi2(1, N=145)=23.891$, p<0.001) (*Table 5).* In the OT group, a vertical deviation alone was predominantly present (24.5%), followed by a combination of exotropia and vertical deviation (20.8%), and a combination of esotropia and vertical deviation

(18.9%). These frequencies were higher compared to the non-OT group (4.3%, 3.3%, and 9.8% respectively) (*Table 4*).

Ocular motility

Ocular motility was assessed in 53 patients with OT and 90 patients without (ocular) torticollis. A strong association (V=0.574) was found between abnormalities in ocular motility and OT ($\chi 2(1, N=143)=47.123$, $p<0.001$) (*Table 5*). Elevation in adduction in one or both eyes was the most frequently observed abnormality in both groups: one eye in 55.3% (n=27) in the OT-group and 21.1% (n=19) in the NT-group, and both eyes in 26.4% (n=14) in the OT-group and 10% (n=9) in the NT-group. In 55% of patients, the elevation in adduction was on the side of the fused suture.

Alphabetical pattern

A significant association was found between alphabetical patterns and OT ($\chi 2(1, n=143) =26.157$, $p<0.01$) with a moderate effect size (V=0.428) *(Table 5)*. In the OT-group (n=53), 54.7% of patients showed a V-pattern and 13.2% of patients showed an A-pattern. In contrast, in the NT-group (n=90), 18.9% of patients showed a V-pattern and 5.6% of patients an A-pattern (*Table 4)*.

Cyclotorsion

In total, the cyclotorsion was determined in 53 patients: 30 patients with OT, and 23 patients without (ocular) torticollis. An ophthalmologist anatomically determined cyclotorsion in most cases (52.8%, n=28). In the OT-group, excyclotorsion was observed in the majority of patients (73.3%,), while only 1 patient (3.3%) showed incyclotorsion. In the NT-group, excyclotorsion was also most common (47.8%), with only 3 patients (13%) showing incyclotorsion (*Table 4)*. Since the cyclotorsion in the NT-group was only determined when abnormalities of the motility were observed or prior to strabismus surgery, no valid comparisons could be made between the two groups and no association could be assessed.

Association between the presence of strabismus, ocular motility abnormalities and alphabetical patterns

When determining the associations for both groups together (n=146), significant associations were found between the presence of strabismus and ocular motility abnormalities ($\chi 2(1, n=142)=33.157$, $p<0.01$), strabismus and alphabetical patterns ($\chi 2(1, n=142)=24.521$, $p<0.01$), as well as between ocular motility abnormalities and alphabetical patterns ($\chi 2(1, n=143)=56.051$, $p<0.01$) *(Table 6)*. So, a logistic regression model could not be conducted due to multicollinearity.

Table 6: Significant associations between predictor variables for the total group of patients included in the study (n=146) were found, indicating multicollinearity. Therefore, determining the individual effects of each variable is impossible in a logistic regression model.

	Presence of strabismus [p-value, Cramers' V]	**Ocular motility abnormalities** [p-value, Cramers' V]	**Alphabetical patterns** [p-value, Cramers' V]
Presence of strabismus	-	<0.01 0.483	<0.01 0.416
Ocular motility abnormalities	<0.01 0.483	-	<0.01 0.626
Alphabetical patterns	<0.01 0.416	<0.01 0.626	-

Refractive error

The refractive error was obtained in 125 patients: 45 patients with OT and 80 patients without (ocular) torticollis. In both the OT-group and NT-group, hypermetropia was most prevalent (89.1% and 89.9, respectively), followed by astigmatism (43.5% and 44.3%, respectively) and anisometropia (26.7% and 26.3%, respectively) *(Table 4).* No significant associations were present between any type of refractive error and OT (p=1.0) (*Table 5)*.

Amblyopia

Amblyopia was present in 33 (61.1%) patients with OT and 31 (33.7%) patients without (ocular) torticollis *(Table 4).* A significant association was found between OT and the presence of amblyopia ($\chi 2(1, n=146) = 10.38$, $p=0.002$), although its effect size was small (V=0.267) *(Table 5).*

Discussion

This study presents the largest cohort of UCS patients compared to the current literature to determine the prevalence of OT and its associated ophthalmic features. In 95% of cases, torticollis in UCS patients is ocular-related. Overall, one-in-three patients with UCS have OT. A significant association was found between the presence of OT and strabismus, ocular motility abnormalities, alphabetical patterns, and amblyopia. UCS patients with OT had a relatively poorer BSV compared to those without torticollis. An excyclotorsion of one or both eyes was present in half of the patients. The findings of this study imply that in patients with UCS, torticollis is highly associated with an underlying ocular cause.

Strabismus is the leading cause of OT (80%) in the general population, with incomitant strabismus as the most frequent form (80%)[47–50]. Nystagmus is found as the second leading cause[16,47–51] followed by infantile esotropia[47,49]. The present study, only including patients with UCS, yielded similar results: strabismus accounted for the majority (82%) of OT cases, with incomitant strabismus (88.7%) being the most prevalent identifiable cause. Only one patient (1.8%) had OT caused by congenital nystagmus. Earlier studies have reported a prevalence of strabismus in UCS patients ranging from 40-70%[22–27]. An incomitant type of strabismus[26,52] was more frequently observed than a concomitant type of strabismus[27,53]. In the present study, a similar occurrence of strabismus was found (47.2%), mostly incomitant. However, within the study groups, the prevalence of strabismus was higher in the OT-group (73.3%) and lower in the NT-group (31.2%). The present study's findings indicate that similarly to the general population, (incomitant) strabismus is the primary cause of OT in patients with UCS.

A reason for adopting OT in (incomitant) strabismus is to achieve BSV, by adjusting their head posture in the gaze direction where the angle of strabismus is least pronounced[17–20.] Surprisingly, no significant association was found between BSV and OT in patients with UCS in this study. Hence, a strong correlation was observed between OT and ocular motility abnormalities, and a moderate association regarding alphabetical patterns. The observed ocular motility abnormalities and alphabetical patterns are consistent with previous studies. Touzé et al.[11,31] demonstrated similar findings in UCS patients: an elevation in adduction was present in 65% (n=13) of patients and a V-pattern strabismus was present in 45% (n=9) of patients. MacIntosh et al.[26] demonstrated an elevation in adduction in half of their included UCS patients. These findings suggest that UCS patients adopt OT in order to overcome ocular motility abnormalities and/or alphabetical patterns in (incomitant) strabismus, and not solely for obtaining BSV.

Ocular manifestations like strabismus, ocular motility abnormalities, cyclotorsion, and amblyopia in UCS are believed to result from various mechanical processes. UCS patients typically exhibit structural differences, including a taller, narrower, and smaller ipsilateral orbit compared to the vertically shorter, wider, and larger contralateral orbit[54,55]. This anatomical variation leads to a shorter and weaker ipsilateral superior oblique muscle[54]. Furthermore, displacement or absence of the superior oblique muscle has been observed[56]. These factors create an imbalance between the inferior- and superior oblique muscle, resulting in a pseudo-superior oblique palsy. Consequently, the inferior oblique muscle overcompensates excessively, leading to vertical manifest strabismus ipsilateral to the side of the synostosis, elevation in adduction, the presence of a V-pattern, excyclotorsion of the eye[28,57] and amblyopia[58,59]. The present study confirms these ophthalmic features in UCS patients and indicates their heightened severity in those with OT. Although a direct association between the presence of OT and cyclotorsion could not be established, excyclotorsion was most commonly observed in both groups. An alternative reason for these more severe abnormal manifestations in UCS patients with OT could be the relatively poorer and more frequent absence of BSV. Previous research supports this hypothesis, demonstrating a strong correlation between the absence of BSV and the occurrence of elevation adduction, V pattern, and excyclotorsion, all of which increased when BSV was absent[60].

Torticollis in patients with UCS is predominantly ocular-related, as demonstrated by the findings of this study, which is important knowledge for plastic surgeons treating these patients. It highlights the importance of timely referral of all UCS patients with torticollis to an orthoptist for screening before considering physiotherapy. Orthoptists are experts in diagnosing OT and identifying the associated ophthalmic features found in these patients, being strabismus, ocular motility abnormalities, alphabetical patterns, amblyopia, and relatively poorer BSV. Orthoptists can effectively treat OT by using glasses or performing strabismus surgery[61]. Timely treatment of OT can prevent complications associated with permanent torticollis including neck pain, headache, muscular and soft tissue changes, facial asymmetry, and scoliosis[16,51,61,62].

The current study's strength is its substantial sample size of 146 patients, exceeding previous studies on ophthalmic features in UCS patients ranging from 15 to a maximum of 59 included patients[11,26,31,56]. Furthermore, this study is the first to differentiate between the presence of these features in cases with OT and those without (ocular) torticollis. However, a limitation of this retrospective study is the absence of protocolized examinations, potentially introducing information bias. The data used in this study was obtained from routine ophthalmic and orthoptic care, lacking protocolized assessments. In routine care, the presence of manifest strabismus often leads to the assumption of absent BSV resulting in a limited number of patients with BSV measurements. Similarly, determining cyclotorsion in patients without torticollis was lacking, as it was primarily assessed in patients preparing for strabismus surgery. Therefore, no valid association between OT and cyclotorsion could be established. We assume that if cyclotorsion was determined in all UCS patients without torticollis, an association between the presence of OT and cyclotorsion would be found. Future studies should consider larger prospective multicenter studies, also including other forms of synostosis creating asymmetry of the skull, with standardized ophthalmological and orthoptic assessments, as well as long-term follow-up evaluations.

Conclusion and recommendations

Torticollis in patients with UCS is predominantly ocular-related. Overall, one-in-three patients with UCS have OT. This makes it essential for plastic surgeons to refer all UCS patients with torticollis to an orthoptist, who possesses the expertise to diagnose and treat OT and its related ophthalmic features. The study's findings suggest a need to reconsider current guidelines for ophthalmic care in UCS patients, placing emphasis on referring all UCS patients with torticollis to an orthoptist for screening before considering physiotherapy.

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