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The turricephaly index: A validated method for recording turricephaly and its natural history in Apert syndrome☆

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ABSTRACT

Introduction: We present the CT scan-derived turricephaly index (TI) as a quotient of the maximal occipito-frontal length of the skull to the distance from the centre of the sella to the highest point on the vertex as a validated tool for assessing turricephaly and evaluating surgical techniques aimed at reducing it.

Materials and methods: Measurements taken from CTs of non-operated children with Apert syndrome and age-matched controls were analysed using *Centricity PACS* system (from the lateral scout image) and the thick-sliced *Osirix* tool. CTs from non-operated children with Apert syndrome were used to investigate the natural history of their turricephaly both as a group and individually.

Results: There was statistically significant agreement between measurements taken from the CT scout and *Osirix* for 42 control children ($R^2 = 0.97$) and 42 children with Apert syndrome ($R^2 = 0.98$) and between two separate observers. There was a statistically significant difference ($p < 0.001$) between CT scout-derived TI value between controls (1.73 ± 0.12, range 1.46–1.99) and Apert children (1.42 ± 0.15, range 1.13–1.73). Analysis of 113 CTs of 65 non-operated children with Apert syndrome showed a decrease in turricephaly with age (positive spearman correlation: $r = 0.50$, $p < 0.001$). Analysis of 37 CTs of those with multiple (>2) CT's showed a similar decrease in turricephaly in the individual child ($p < 0.001$).

Conclusions: TI derived from the CT scout view provides a simple, objective and validated method for assessing turricephaly. We recommend it for monitoring and for the prospective evaluation of reconstructive techniques in children with complex/syndromic craniosynostosis.

1. Introduction

Turricephaly (*Tower Head*) is a descriptive term for a distinctive head shape often observed in children with syndromic forms of craniosynostosis, in particular, those with Apert syndrome (Ferguson, 1980). As an anthropometric measurement it has been poorly defined[1] and this has resulted in craniofacial surgeons relying on subjective assessments when describing its severity and when evaluating both its natural history and the effects of surgical intervention (Sonstein et al., 1996; Weber et al., 2008). Quotients of anthropometric measurements of skull dimension are already used to describe certain head shapes (the cranial index – CI – in scaphocephaly, for example) but vertical height has been omitted because reliable reference points can be difficult to define (Waitzman et al., 1992b).

There is therefore a need for a simple, objective and validated method for assessing turricephaly similar to the CI that is now frequently used as a proxy measure for scaphocephaly (Waitzman et al., 1992b; Fearon et al., 2006). CI is simple to derive and can be used both to record the severity of scaphocephaly and to compare the effectiveness of surgical techniques used to reduce it (Nowinski et al., 2012; Saiepour et al., 2013; Derderian et al., 2012; Goldstein

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et al., 2013). The turricephaly index (TI) is, like the CI, a quotient derived from two simple radiologically determined measurements but substituting vertical height (Waitzman et al., 1992a) for width.

To be accepted, a new index should be validated for the methodology used, for inter-observer variation and for its clinical utility. This study deals with all these issues. In Part 1 we have calculated TI using two different imaging techniques – one simple, one more complex and (a) compared the results obtained from each; (b) recorded consistency between observers and (c) compared TI of children with Apert syndrome with those of normal controls. In Part 2, the clinical utility of TI has been tested on a predictably affected model – the child with Apert syndrome – to (a) examine variations in TI with age in a cohort of non-operated children with Apert syndrome; and (b) investigate variations in TI in the individual child with Apert syndrome.

2. Materials and methods

The retrospective review of patient data for this study was approved by the hospital's research and audit department (audit registration number 1544).

For Part 1 of the study, CT scans of patients with Apert syndrome presenting to the craniofacial unit at Great Ormond Street Hospital for Sick Children, London, UK between 2000 and 2017 were retrieved from the craniofacial unit database. Age- and sex-matched controls who had CT scans (on the same equipment and at the same settings) for computer navigation-guided epilepsy surgery planning were recruited from the neurosurgical unit database. No control patient had conditions affecting their intracranial pressure or head shape.

For Part 2, non-operated children with Apert syndrome who had undergone multiple CT scans were selected.

Spiral CT scans were carried out on a Somatom Sensation Spiral computed tomographic scanner, Siemens, Munich, Germany with 0.75 mm collimation (slice thickness). Standardization of head position was maintained by established protocol[4] of laser light positioning, position maintenance with restraints and optimization of gantry. Scan data were stored as Digital Imaging and Communications in Medicine Files (DICOM, Rosslyn, Va. USA) and exported to both Centricity GE Healthcare IT, (Version 3.1.4, United Kingdom) and open-source software *Osirix* v5.8 (Pixmeo, Geneva, Switzerland).

Scan data in *Osirix* was viewed in 3D multiplanar reconstructions (MPR) with the thick-slab tool set to the maximum of 100 mm (giving a composite view of 100 mm depth in each plane), in the maximum intensity projection setting.

In the sagittal CT scout measurement method the maximal occipito-frontal length (OFL) of the skull (from outer cortex to outer cortex regardless of its plane relative to the skull base) and the distance from the centre of the sella to the highest point on the vertex (SVD) were measured on the calibrated scout image using the calibrated line ruler tool (Figs. 1 and 2).

As a validating comparator, accurate measurements were made of the true greatest OFL (again regardless of the plane upon which it lay) and SVD using *Osirix*. 3D MPRs were viewed (in all three planes) to identify and mark the sella, vertex and the most projected frontal and occipital points. These reconstructions were then re-sliced though the coronal and axial planes on which these points lay, once to identify the OFL plane and once to identify the SVD plane, as viewed sagittally. These distances were measured using the ruler tool (Figs. 3–5).

All measurements were repeated twice and performed independently by two investigators (JOH, BW) at separate times. The data was entered into a spreadsheet and TIs calculated as OFL:SVD for all scans. Statistical analysis was undertaken in *SPSS* (*IBM Corp*., *NY*) and *Matlab* (*MathWorks*, *MA*).

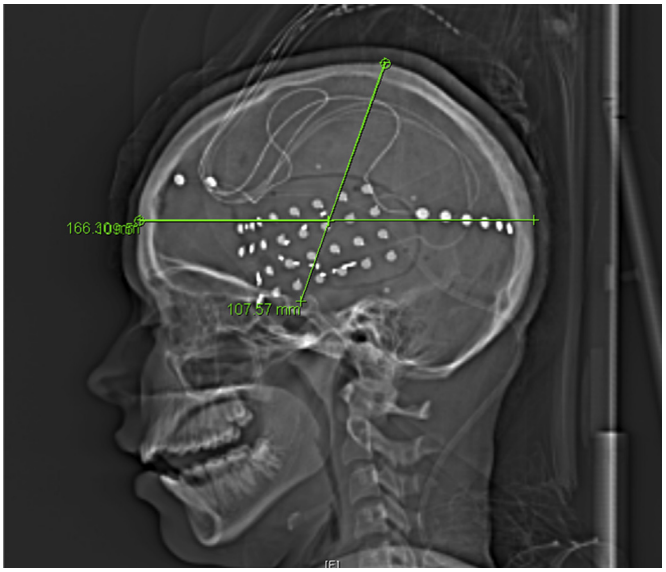

Fig. 1. Measurements used to calculate TI from CT scout image in a "control" child.

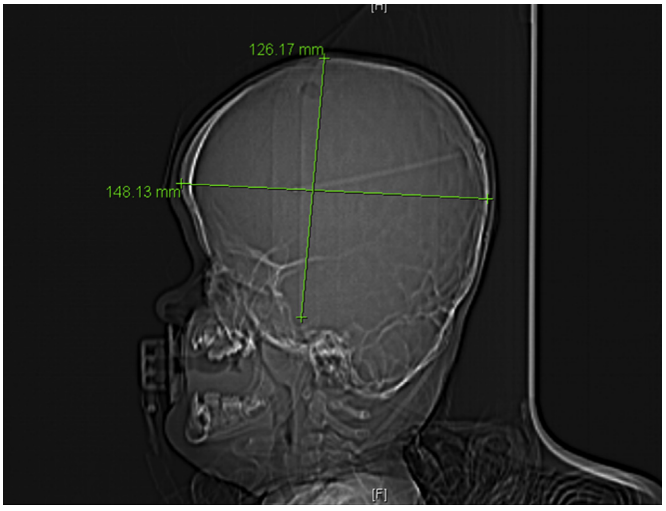

Fig. 2. Measurements used to calculate TI from CT scout image in a child with Apert syndrome.

2.1. Statistical analysis

Intraclass Correlation Coefficient (ICC) and Pearson correlation coefficients were used to compare the TI measurements performed using CT scout vs. Osirix as well as by different observers. Pearson correlation was used to assess age dependence of TI in the Apert population. One-way ANOVA was used to compare mean TI in the three subgroups; paired T-test was used to assess difference between the TI at the time of first CT with that at the time of the last CT available for a subgroup of Apert patients. The Shapiro–Wilk test was used to assess normality of distributions.

3. Results

3.1. Part 1 (Validation)

The CT scans of 42 children with Apert syndrome (average age 8.5 years, range 0.05–20.6 years) and of 42 age and sex-matched

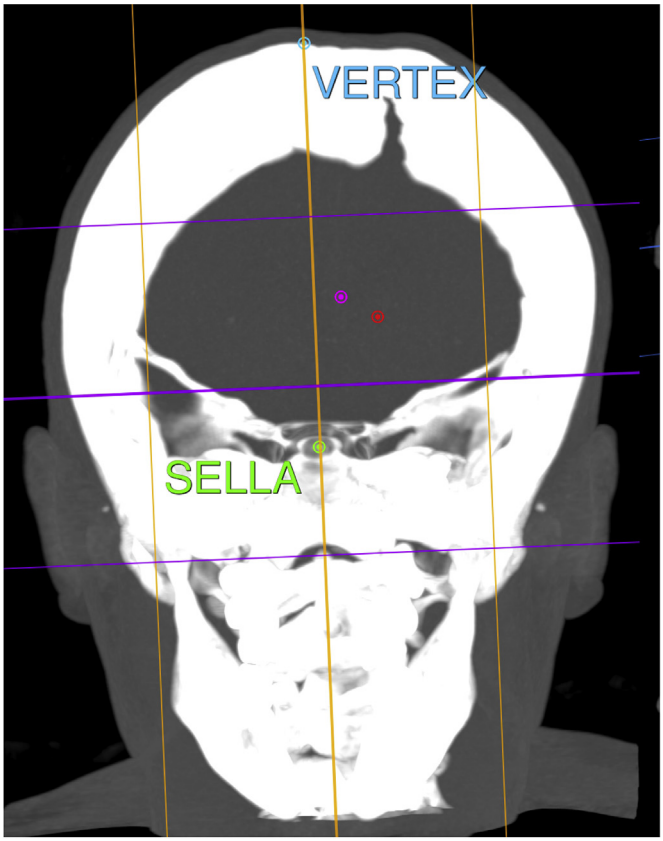

Fig. 3. Coronal 3D MPR re-sliced through sella and vertex on *Osirix*.

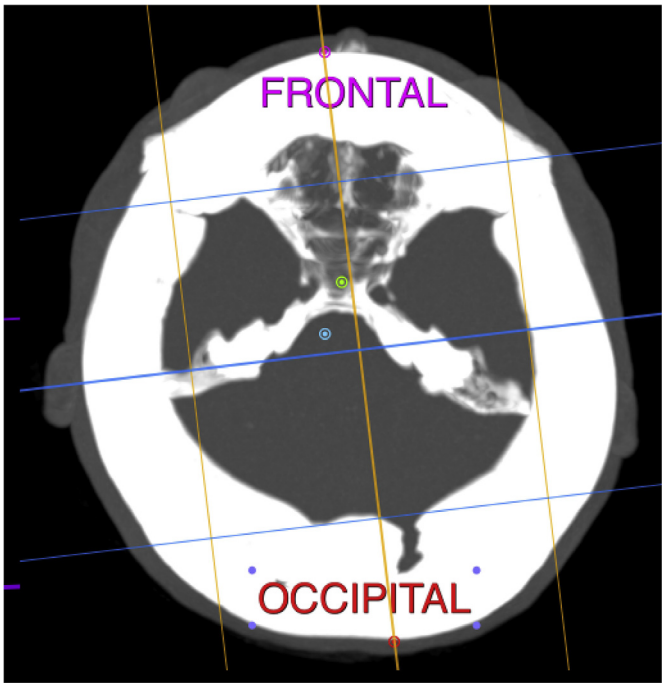

Fig. 4. Axial 3D MPR re-sliced through most projected frontal and occipital points on *Osirix*.

controls (average age 9.9 years, range 1.7–17.8 years) were analysed for this part of the study.

Comparison of the two methods of measurements showed extremely good correlation between those performed on CT scout and Osirix both for control subjects ($R^2 = 0.969$, $p < 0.001$) and Apert patients ($R^2 = 0.994$, $p < 0.001$) (Fig. 6).

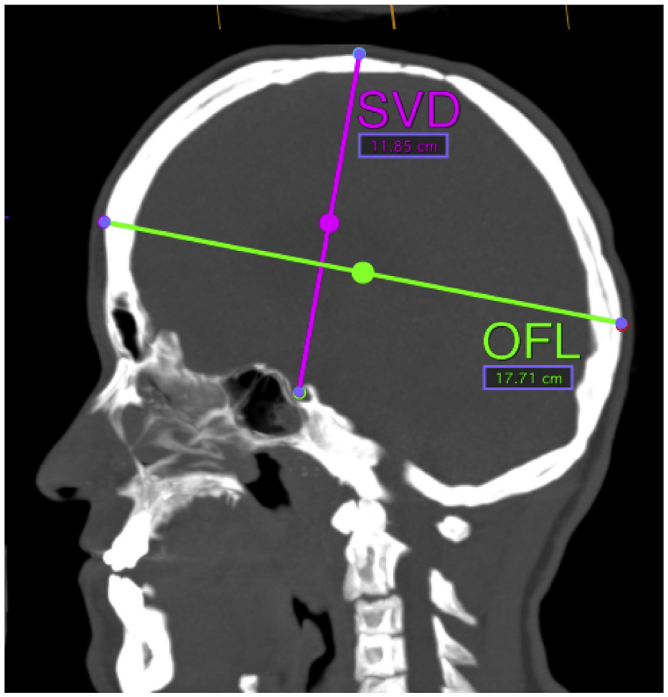

Fig. 5. Composite SVD and OFL measurements on sagittal view on *Osirix*.

OFL and SVD measurements performed by two separate observers showed excellent correlation (Intraclass Correlation coefficient – ICC – above 0.9) when measured on the CT scout view and when using Osirix (Table 1).

The mean TIs derived from these measurements by the CT scout method were 1.42 ± 0.15 for children with Apert syndrome (range 1.13–1.73) and for the controls 1.73 ± 0.12 (range 1.46–1.99)– a statistically significant difference ($p < 0.001$). Fig. 7 shows TI versus age distribution for the two populations.

3.2. Part 2 (Clinical application)

To investigate variations in TI with age, 113 CT scans of 65 Apert patients (average age at scans 4.29 years, range 0–18 years) who had undergone neither cranial vault surgery nor CSF diversion were analysed after the population had been subdivided into three clinical age groups. TI's ranged from 1.26 ± 0.11 (for children <1 year of age) to 1.41 ± 0.16 (for children >5 years of age) (Fig. 8). Table 2 reports all values for each subgroup.

One-way ANOVA showed significant difference between the three subgroups ($p < 0.001$). TI increased with age as shown by positive spearman correlation ($r = 0.50$, $p < 0.001$) (Fig. 9).

To investigate changes in TI in the individual non-operated Apert child the CTs of 37 patients who had undergone more than one CT (number 2–3) with no intervening cranial surgery were analysed. There was a statistical increase in TI from first to last scan ($p < 0.001$) as shown in Fig. 10.

4. Discussion

Turricephaly, despite often responsible for a major component of the craniofacial deformity affecting children with syndromic/complex forms of craniosynostosis, has proven difficult to quantify reliably (Sonstein et al., 1996).

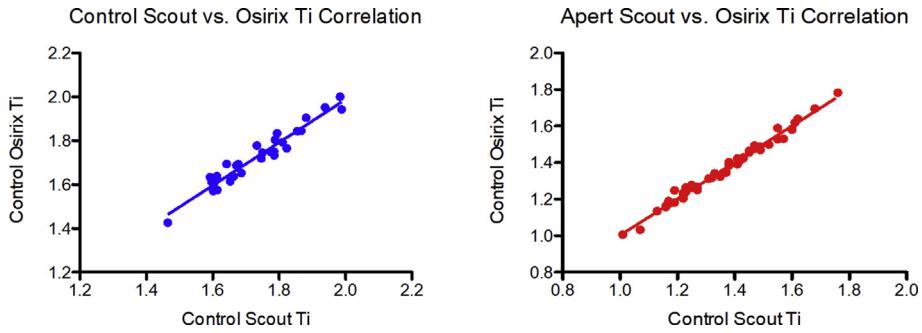

Fig. 6. Correlation between TI measurements performed on *Osirix* and extracted from CT scout for the control group (left) and Apert group (right).

Table 1
ICC of OFL and SVD for measurements performed on Scout and Osirix.

Measured distance	Scout (ICC; p-value)	Osirix (ICC; p-value)
OFL	0.962 ($p < 0.0001$)	0.929 ($p < 0.0001$)
SVD	0.943 ($p < 0.0001$)	0.871 ($p < 0.0001$)

ICC = Intraclass Correlation Coefficient; OFL = Occipito-Frontal Length; SVD = distance from the centre of the sella to the highest point on the vertex.

Table 2
TI value for the different subpopulations.

Age	Apert		
	n	MEAN	SD
<1	55	1.26	0.11
1 to 5	34	1.37	0.11
>5	25	1.41	0.16

TI = Turricephaly Index.

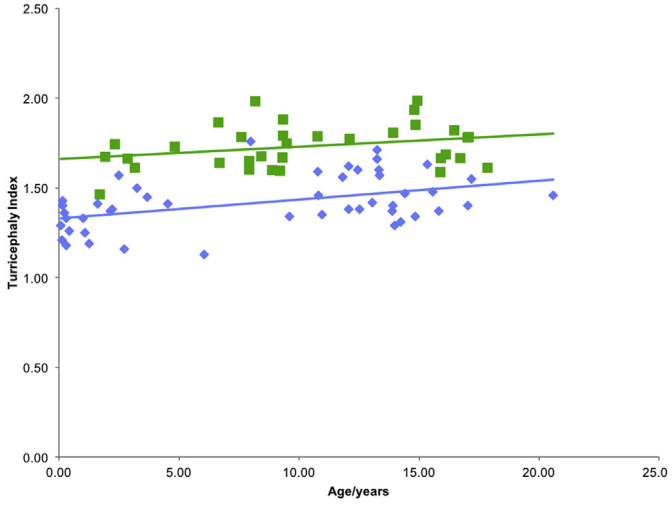

Fig. 7. Age distribution for the Apert and control populations.

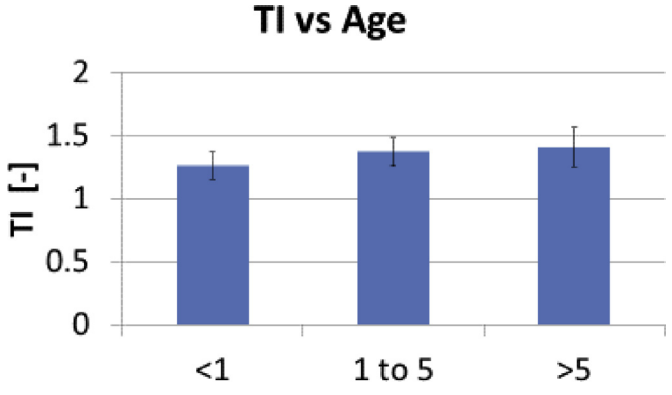

Fig. 8. Average TI values for three subpopulations: younger than 1 year old; between 1 and 5 year old; over 5 year old.

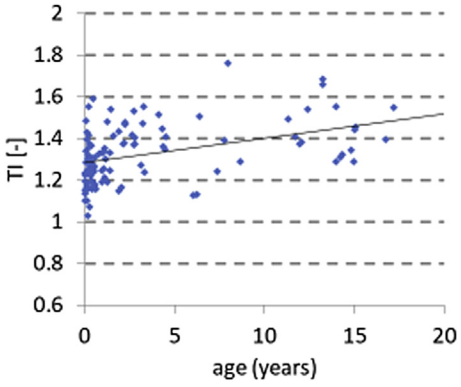

Fig. 9. TI increase with age ($r = 0.5$, $p < 0.001$).

To address this issue we have derived a Turricephaly Index (TI) from simple measurements taken from the lateral scout view of a CT.

As evidence for its validity as a reliable tool for the craniofacial surgeon we have:

- Demonstrated that measurements taken from the scout view of a CT scan are as reliable as those derived from a more complex and precise tool – *Osirix*.
- Established from separate measurements made at separate times (and repeated) by two observers the inter-observer consistency of these CT scan derived measurements.

We submit that the TI so derived provides the craniofacial surgeon with a useful tool for assessing and comparing the effectiveness of surgical maneuvers designed to address a complex skull deformity.

We would not suggest that TI describes the head shape of the affected child in anything but simple terms. The deformity of the child with sagittal synostosis is often more complex than scaphocephaly alone – a description that omits the pterional indrawing and low posterior vertex important in the deformity. Similarly, the head shape of the child with Apert syndrome presents an

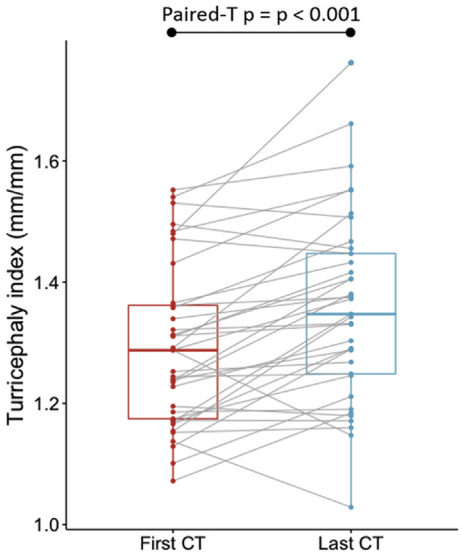

Fig. 10. Turricephaly Index between first and last CT for 37 non-operated patients. Grey lines indicate change for each patient between the first and last CT. Boxplots indicate mean, as well as first and third quartiles.

appearance more complex than turricephaly alone, with its shallow and externally rotated orbits and hypertelorism, for example. Nevertheless, the cranial index (CI) has become an accepted proxy for scaphocephaly within the craniofacial community and has been regularly used for evaluating techniques aimed at restoring a more regular head shape. It is in this context that we propose the TI as a suitable tool for the assessment of turricephaly and evaluation of techniques designed to reduce it.

Every CT scan exposes the patient to ionizing radiation. We are not however suggesting that children with Apert syndrome (for example) should be scanned for the sole purpose of tracking their turricephaly. They are likely in any case to be scanned before and after the various procedures (including CSF diversion) they inevitably undergo in their journey from infancy to adulthood and it is the TI derived from these studies that can be used to judge effect on the child's turricephaly.

Easy-to-derive indices such as the TI and CI can also be combined to provide the craniofacial surgeon with a simpler tool for describing the head shapes of children with craniosynostosis in three rather than two dimensions than those dependent upon more complex analytic techniques such as three-Dimensional handheld scanning and principal component analysis (Rodriguez-Florez et al., 2017; Pluijmers et al., 2012; Staal et al., 2015).

To demonstrate the clinical use of TI we have focused on the turricephaly of children with Apert syndrome and shown how their TI differs from that of age-matched controls (1.42 for children with Apert syndrome versus and 1.73 for controls ($p < 0.001$)).

Although it has been suggested that untreated turricephaly in Apert syndrome is likely to worsen with age (a possible indication for early surgical intervention (Nowinski et al., 2012; Allam et al., 2011)) our study shows the opposite – there is a tendency for the degree of turricephaly to reduce with age and approach more normal proportions (an increase in TI from 1.25 under 1 year old to 1.41 over age 5 years against a mean TI of our control population of 1.73). On this evidence, the craniofacial surgeon should not suggest to parents of affected children that a predictable worsening of turricephaly in the absence of other factors is a valid reason for early reconstructive surgery. Many operations, a posterior vault expansion, for example, Nowinski et al. (2012), Goldstein et al. (2013), Arnaud et al. (2012), De Jong et al. (2013) may indeed reduce turricephaly but, our data shows, such procedures are not indicated for prophylaxis alone.

What accounts for this decline in the severity of turricephaly in Apert syndrome? In the young Apert child the sagittal and metopic sutures are often widely open with the coronals and lambdoids wholly or partially closed – a combination that constricts the skull base in the antero–posterior axis and predisposes to upward expansion – turricephaly. But when, after two or three years, all vault sutures are likely to have fused skull growth becomes more evenly distributed with a comparative reduction in the previous upward drive.

5. Conclusion

We propose the TI as a tool as simple, reliable and repeatable as the CI for use as a proxy when describing one element of the calvarial deformity seen particularly in children with Apert syndrome but also in other complex forms of craniosynostosis. Like the CI for scaphocephaly it can be used both to track the progress of turricephaly in the individual patient and to evaluate the effects of surgical procedures designed to reduce it.

In addition, we have demonstrated how, for the unoperated child with Apert syndrome, there is a statistically significant tendency for this particular proportion of their complex craniofacial deformity to improve with time.

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Conflicts of interest

The authors declare no conflicts of interest.

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