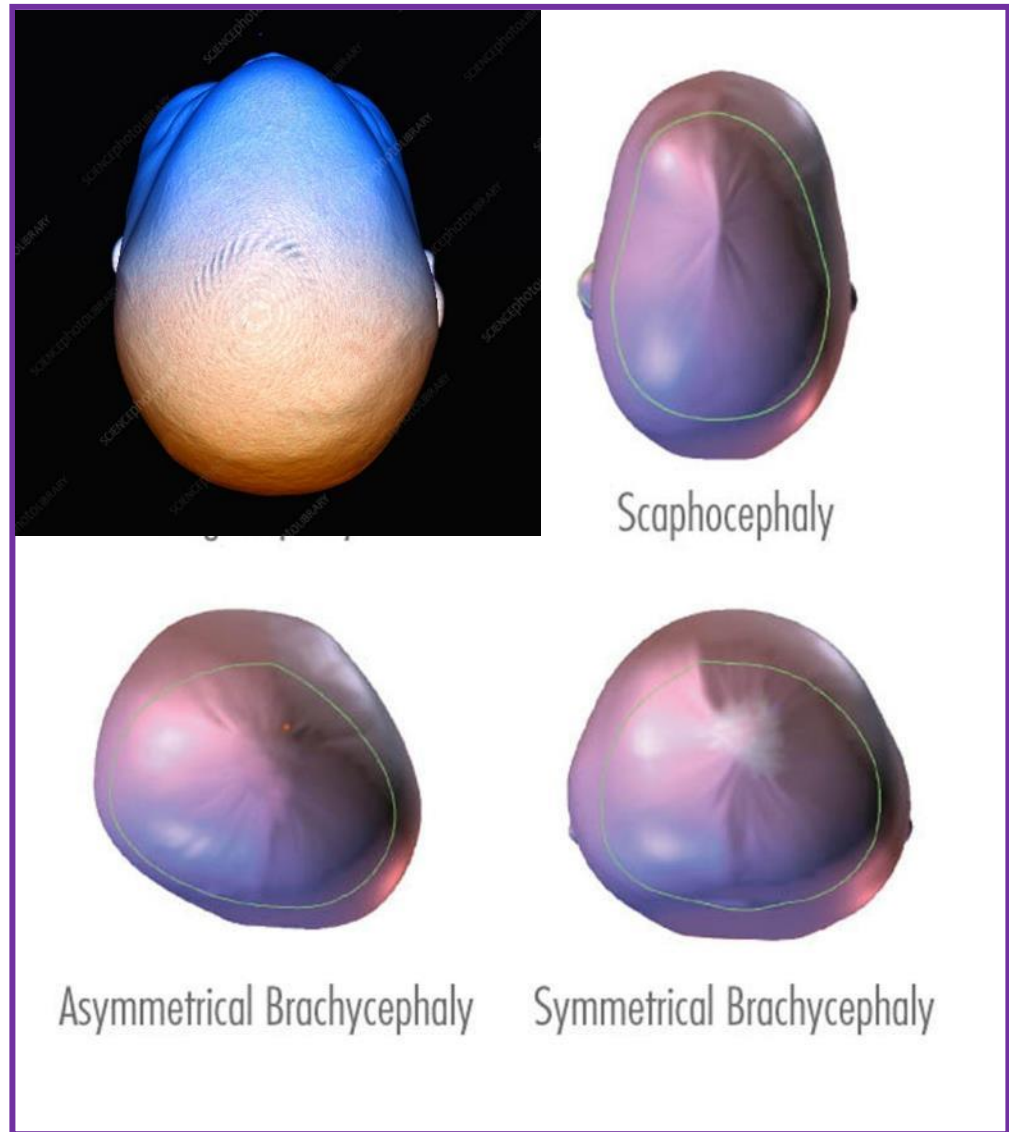


Abnormal skull shape

가톨릭의대 인천성모병원
장대현

Terminology

- Plagiocephaly
- Scaphocephaly (Dolichocephaly)
- Brachycephaly
- Trigonocephaly
- Macrocephaly/microcephaly





Normal



<https://www.cranialtech.com/online-assessment/>

Classification

- Craniosynostosis
- Non-synostotic plagiocephaly (brachycephaly)
(Deformational/Positional)

Anatomy

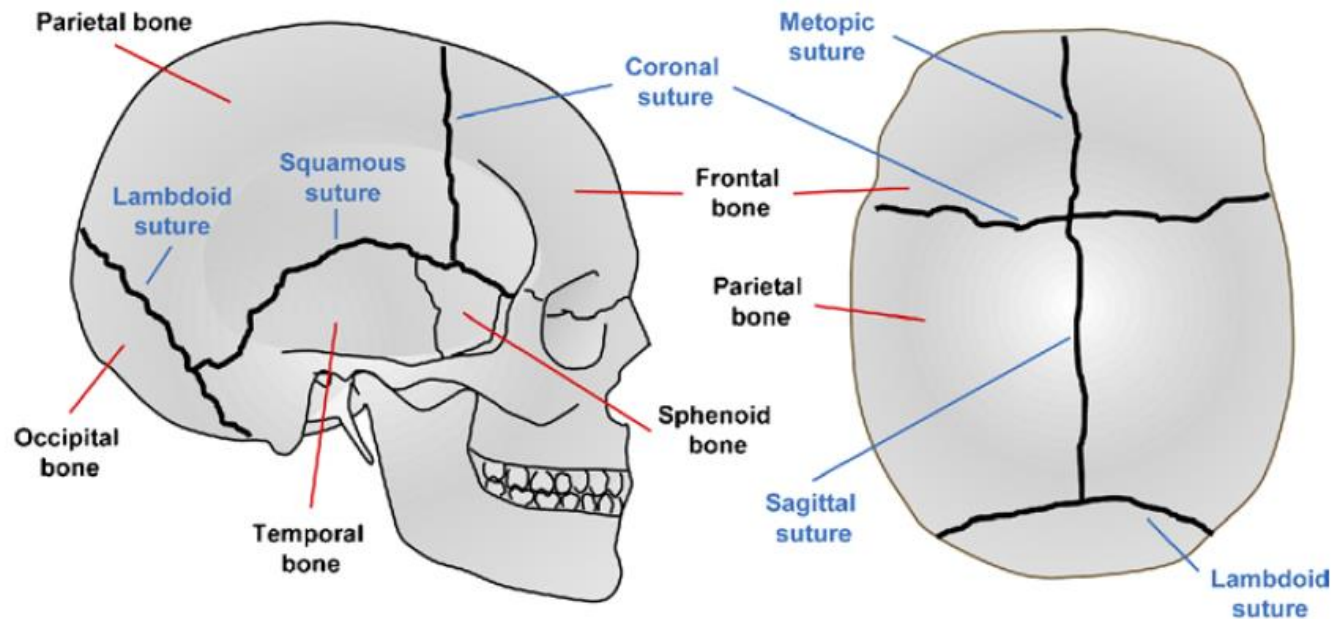
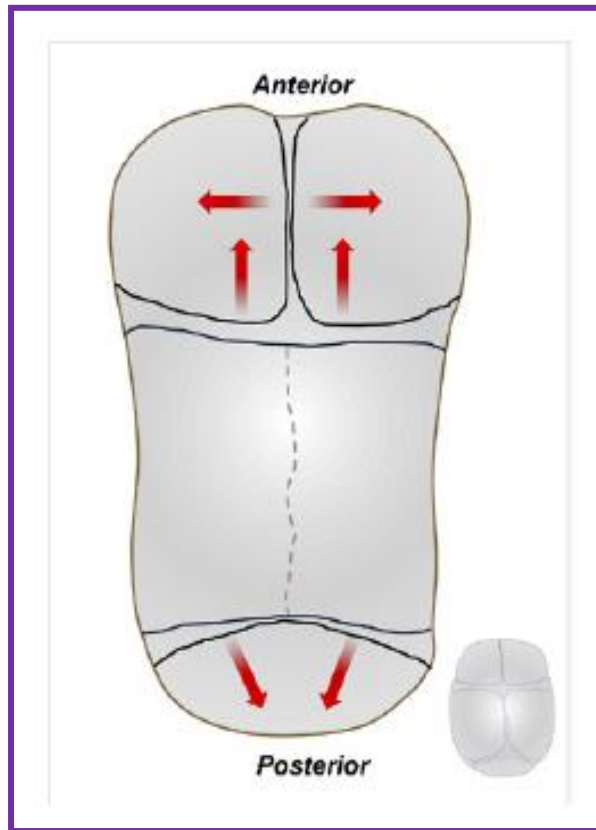


FIGURE 1. Major bones and sutures of the adult human cranium. Lateral (left) and top (right) view demonstrating the bones (red line) and sutures (blue) of the calvarium. The metopic suture separating the right and left halves of the frontal bone generally closes by the second year of life.

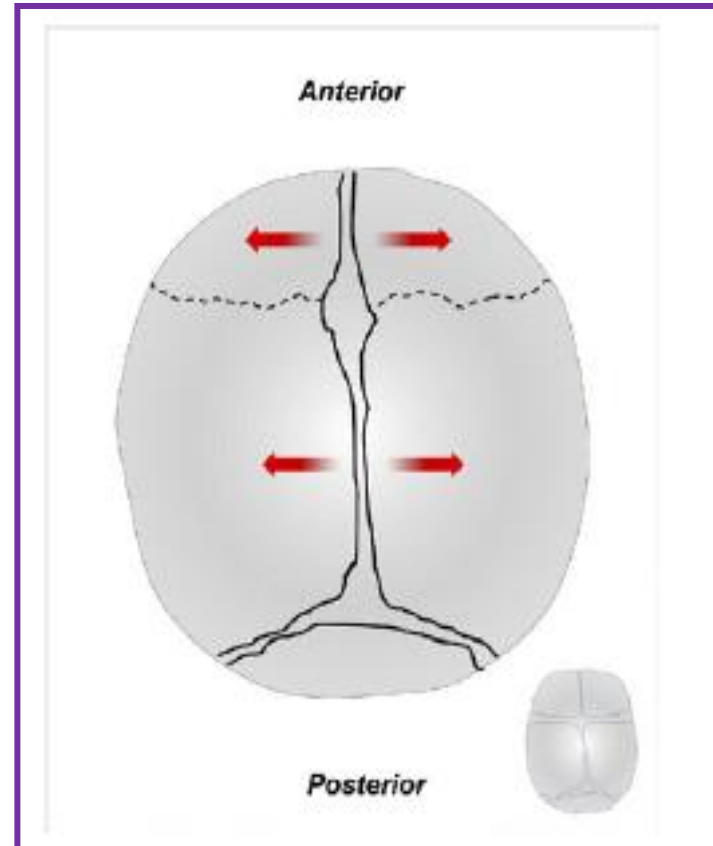
Anatomy

Suture	Timing of fusion
Metopic suture	3–9 months of age
Coronal suture	Between 22 and 39 years of age
Sagittal suture	Between 22 and 39 years of age
Lambdoid suture	Between 22 and 39 years of age

Craniosynostosis

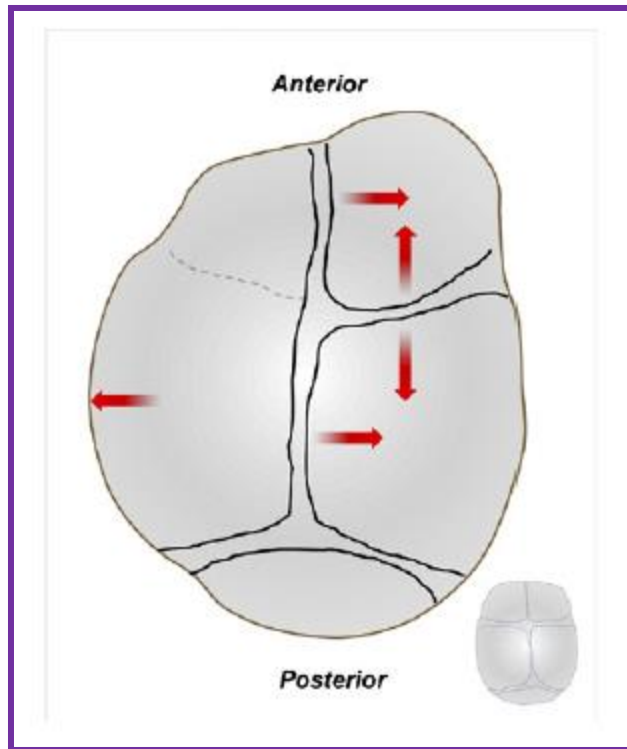


Sagittal synostosis (scaphocephaly).

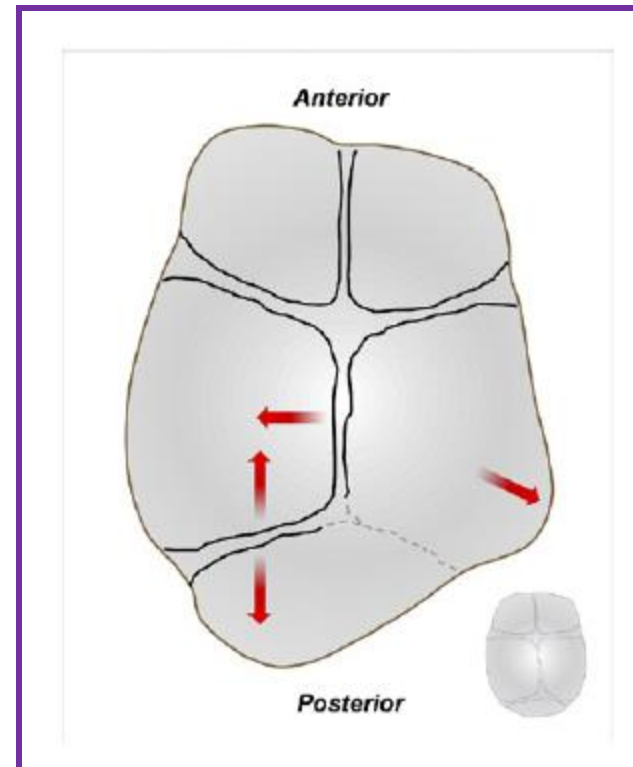


Bilateral coronal craniosynostosis.

Craniosynostosis



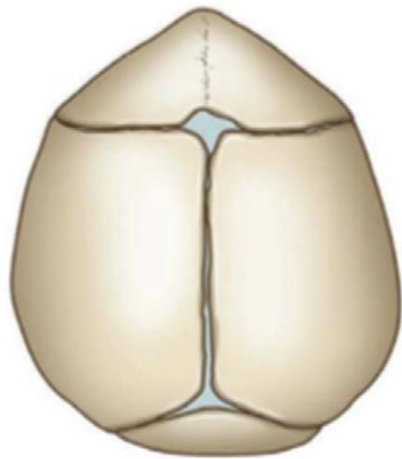
Unilateral coronal craniosynostosis.



Lambdoid craniosynostosis.

Non-synostotic plagiocephaly (brachycephaly) (Deformational/Positional)





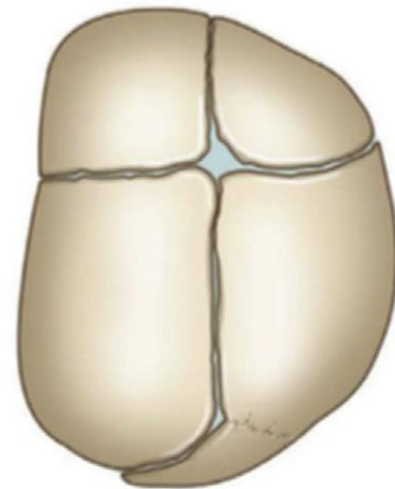
Metopic

Synostotic Trigonocephaly



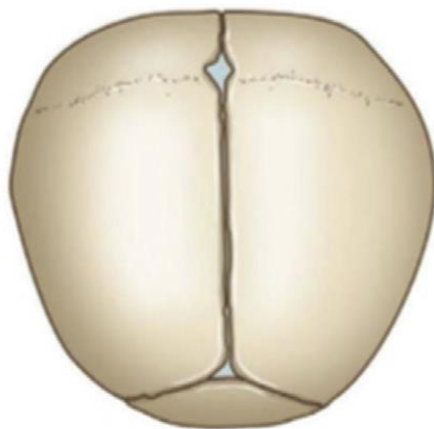
Sagittal

Synostotic Scaphocephaly



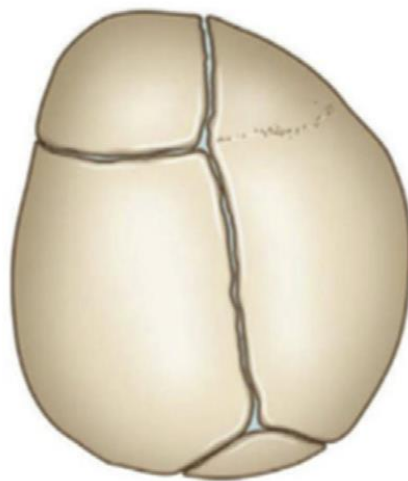
Lambdoid

Synostotic Posterior Plagiocephaly



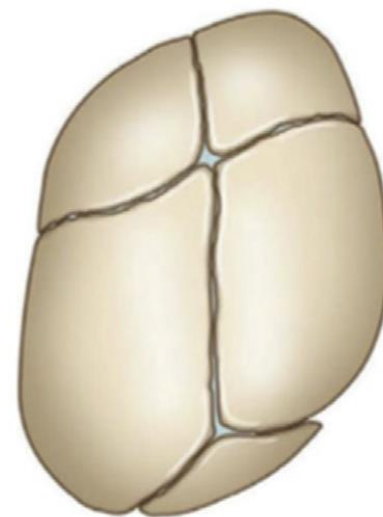
Bicoronal

Synostotic Brachycephaly



Unicoronal

Synostotic Anterior Plagiocephaly



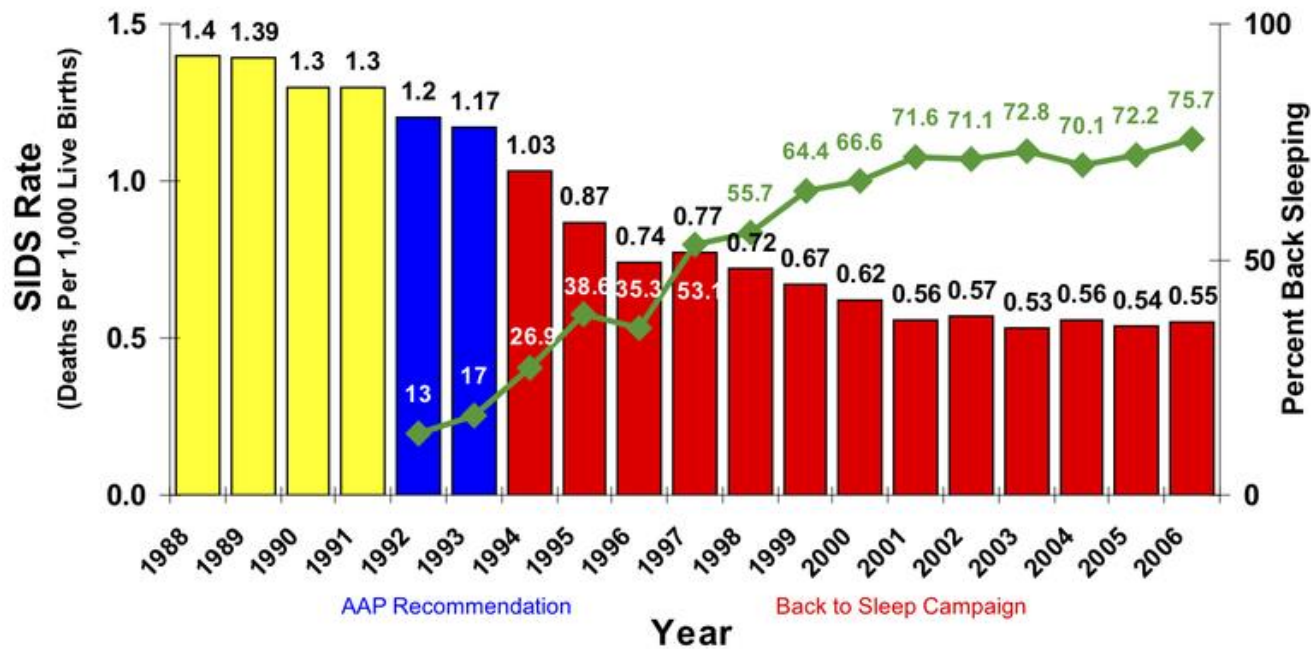
(All Sutures Open)

Deformational Posterior Plagiocephaly

- Craniosynostosis
- Non-synostotic plagiocephaly (brachycephaly)
(Deformational/Positional)

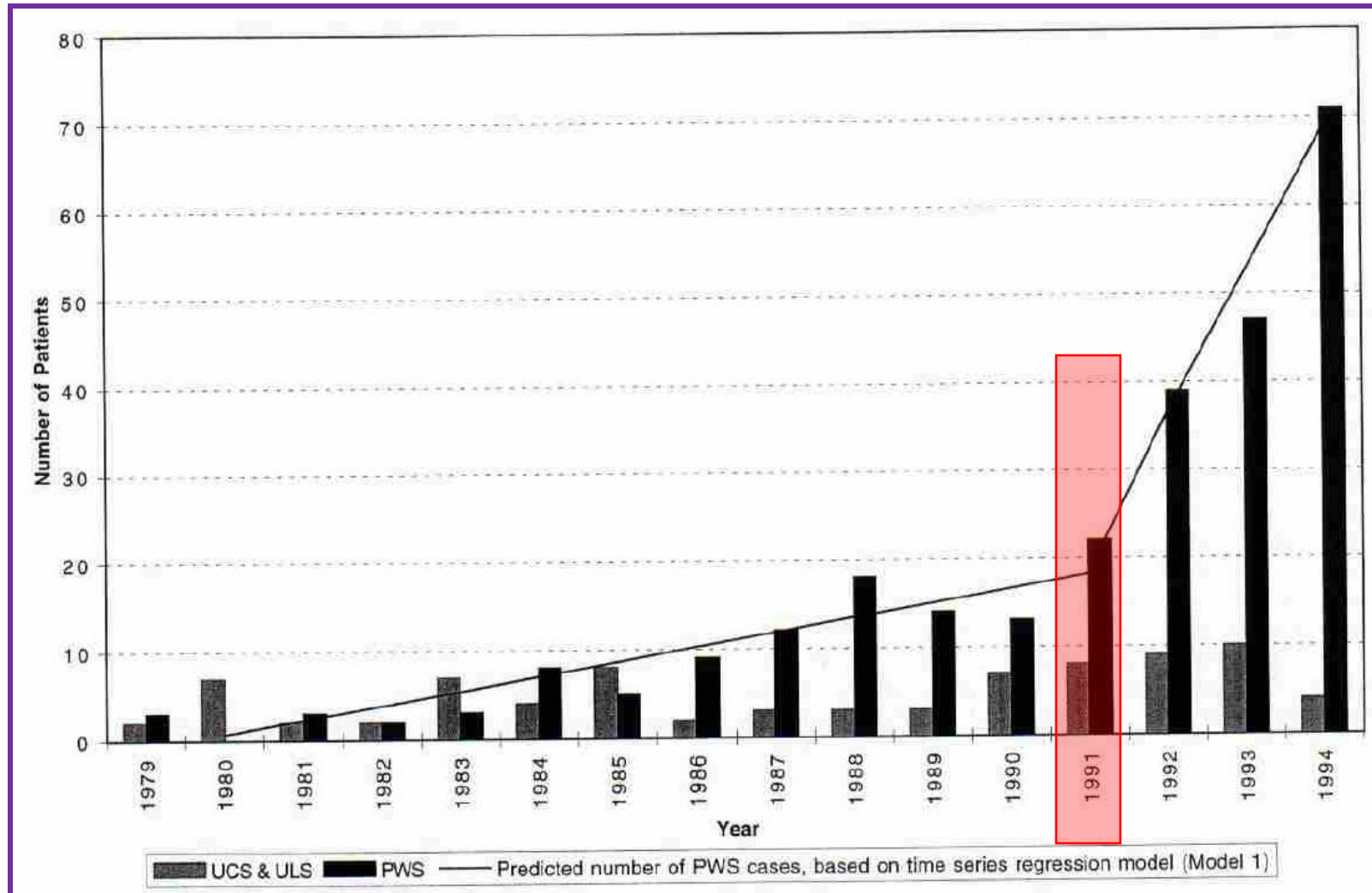
Deformational (Positional) plagiocephaly

SIDS Rate and Back Sleeping (1988 – 2006)



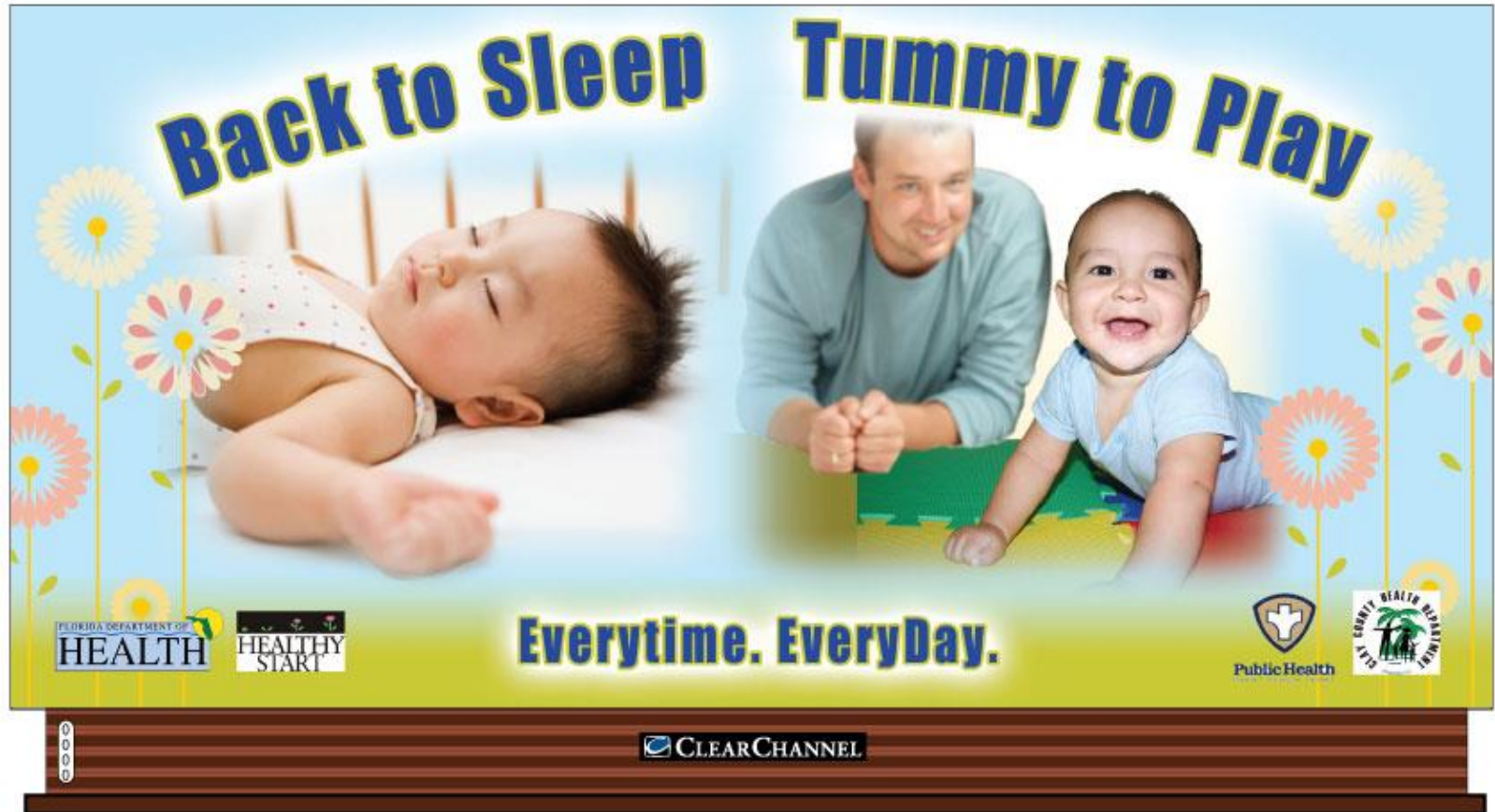
SIDS Rate Source: CDC, National Center for Health Statistics,
Sleep Position Data: NICHD, National Infant Sleep Position Study.

Observations on a Recent Increase in Plagiocephaly Without Synostosis



Tummy to Play

Premier Vinyl 10'9" X 23' Scale 1/2" = 1'



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We will gladly make up to 2 revisions to original design free of charge.

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Additional Revision Date: _____ Total Hours: _____

Art Approval: _____ Date: _____

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Source: _____ Image#: _____ Price: _____

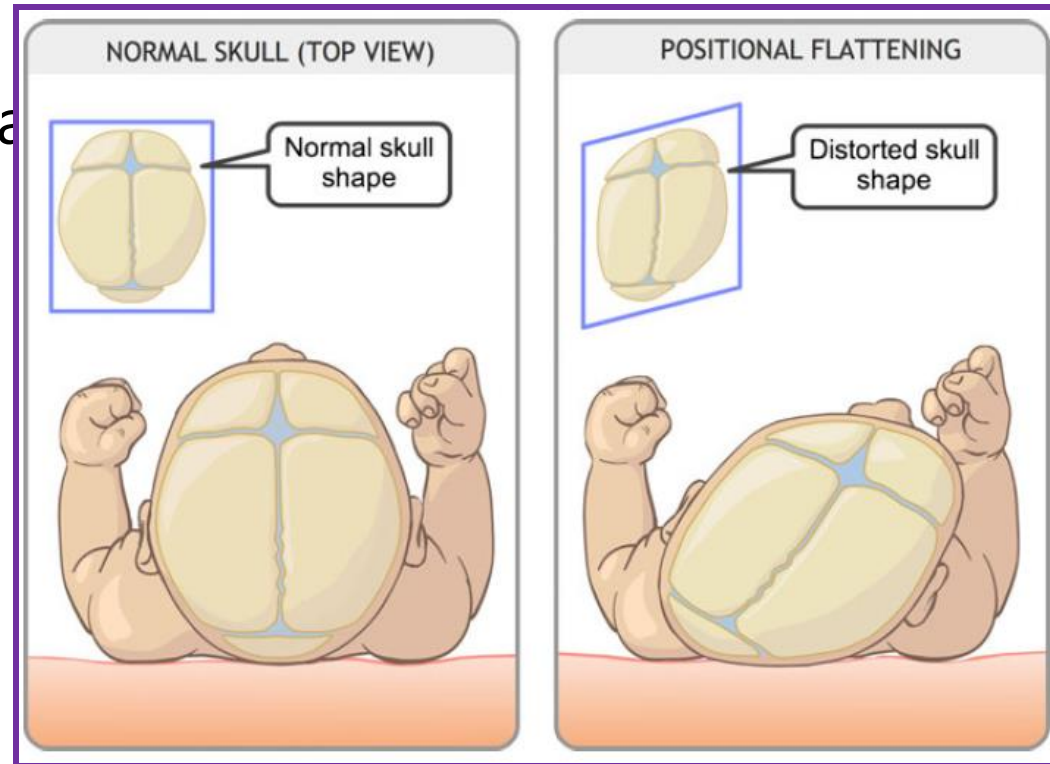
This outdoor design has been created by Clear Channel Outdoor. Reproduction of this display in whole or part without permission, is prohibited.

File: Safe Sleep	Date: 9/2/11
Artist: Brandi	AE: Amber
Extension Sq. Ft.:	Board #:
Approved By:	Date:

Deformational (Positional) plagiocephaly

- Mechanical factors (utero, at birth, postnatal)

- Torticollis
- Prematurity (incubator)
- Twin
- Neuromuscular disease
- Hypotonia
- Sleep position



Plagiocephaly and Brachycephaly in the First Two Years of Life: A Prospective Cohort Study

- 200 infants (1.5, 4, 8, 12, 24 months)

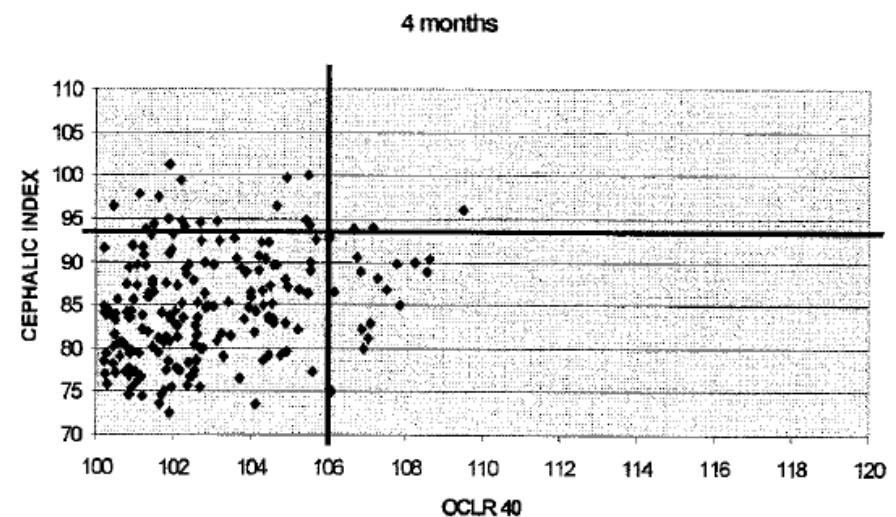
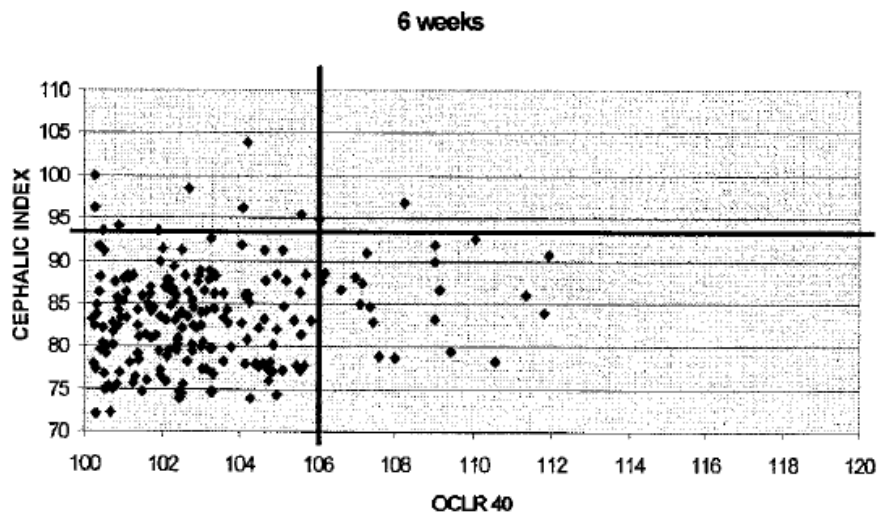







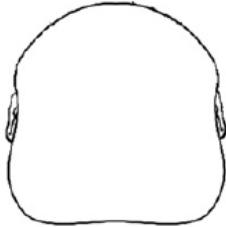
TABLE 5. Significant Risk Factors at 6 Weeks

Variable	Case (<i>n</i> = 32; <i>n</i> [%])	Control (<i>n</i> = 168; <i>n</i> [%])	Univariate OR (95% CI, <i>P</i> Value)	Multivariate OR (95% CI, <i>P</i> Value)
Limitation of passive rotation - newborn (missing = 16)			$\chi^2 = 12.52, P = .0004$	$\chi^2 = 11.50, P = .0007$
Limited	7 (25.0)	8 (5.1)	6.17 (2.03–18.76)	9.51 (2.59–34.94)
Not limited	21 (75.0)	148 (94.9)	1.00	1.00
Reported preferential head orientation (missing = 2)			$\chi^2 = 3.19, P = .07$	
Yes	23 (71.9)	91 (54.8)	2.11 (0.92–4.83)	
No	9 (28.1)	75 (45.2)	1.00	
Sleep position at newborn interview			<i>P</i> < .0001	
Supine only	30 (93.8)	95 (56.5)	11.53 (2.67–49.81)	
Nonsupine	2 (6.2)	73 (43.5)	1.00	
Sleep position at 6 wk			$\chi^2 = 11.79, P = .0006$	$\chi^2 = 9.25, P = .003$
Supine only	26 (81.3)	81 (48.2)	4.65 (1.82–11.89)	5.27 (1.81–15.39)
Nonsupine	6 (18.7)	87 (51.8)	1.00	1.00
Head position varied			$\chi^2 = 10.94, P = .004$	
Yes	10 (31.2)	103 (61.3)	1.00	
No	12 (37.5)	45 (26.8)	2.75 (1.11–6.82)	
Tried but unsuccessful	10 (31.2)	20 (11.9)	5.15 (1.90–13.98)	
Back time per day			$\chi^2 = 8.57, P = .003$	
< 21 h	16 (50.0)	128 (76.2)	1.00	
\geq 21 h	16 (50.0)	40 (23.8)	3.20 (1.47–6.97)	
Upright time per day			$\chi^2 = 4.90, P = .03$	$\chi^2 = 6.88, P = .009$
\leq 1 h	9 (28.1)	83 (49.4)	1.00	1.00
> 1 h	23 (71.9)	85 (50.6)	2.50 (1.08–5.56)	3.99 (1.42–11.23)
Backtime per day (h)	19.01 (4.13)	14.89 (6.52)	<i>P</i> < .0001 $\beta = 0.14 (0.05–0.22)$	

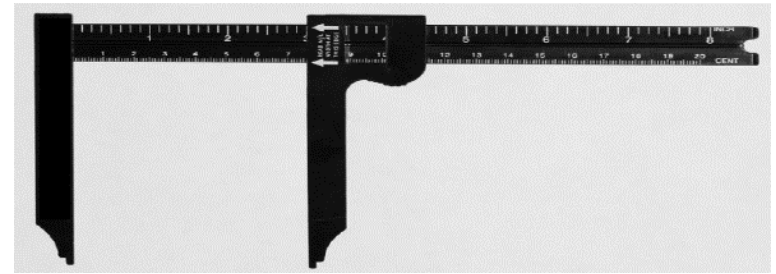
Evaluation

Visual evaluation

FIGURE 1. Vertex views of mild, moderate, and severe lateral and posterior deformational plagiocephaly.

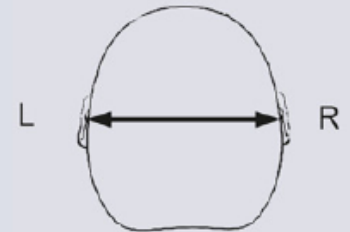
	Mild	Moderate	Severe
Lateral deformational plagiocephaly			
<i>Key findings:</i>	<i>Flattening on back of skull only</i>	<i>Ipsilateral ear anteriorly displaced, ipsilateral frontal bossing</i>	<i>Ipsilateral temporal skull growth</i>
Posterior deformational plagiocephaly (Brachycephaly)			
<i>Key findings:</i>	<i>Central posterior deformity</i>	<i>Widening of posterior skull</i>	<i>Temporal bossing</i>

Measurement



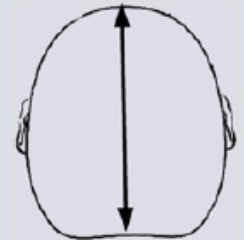
Cranial width (breadth)

The greatest transverse diameter of the head, on a horizontal plane



Cranial length

The distance from the forehead to most posterior point of the head, in the same plane as measured in head circumference



Cephalic index (cranial index)
Occipital-frontal transcranial diameter

The ratio of the cranial width to the cranial length
Determine the sites on the left and right sides of the head where the deformation is the most prominent; measure the diagonal distances between these sites

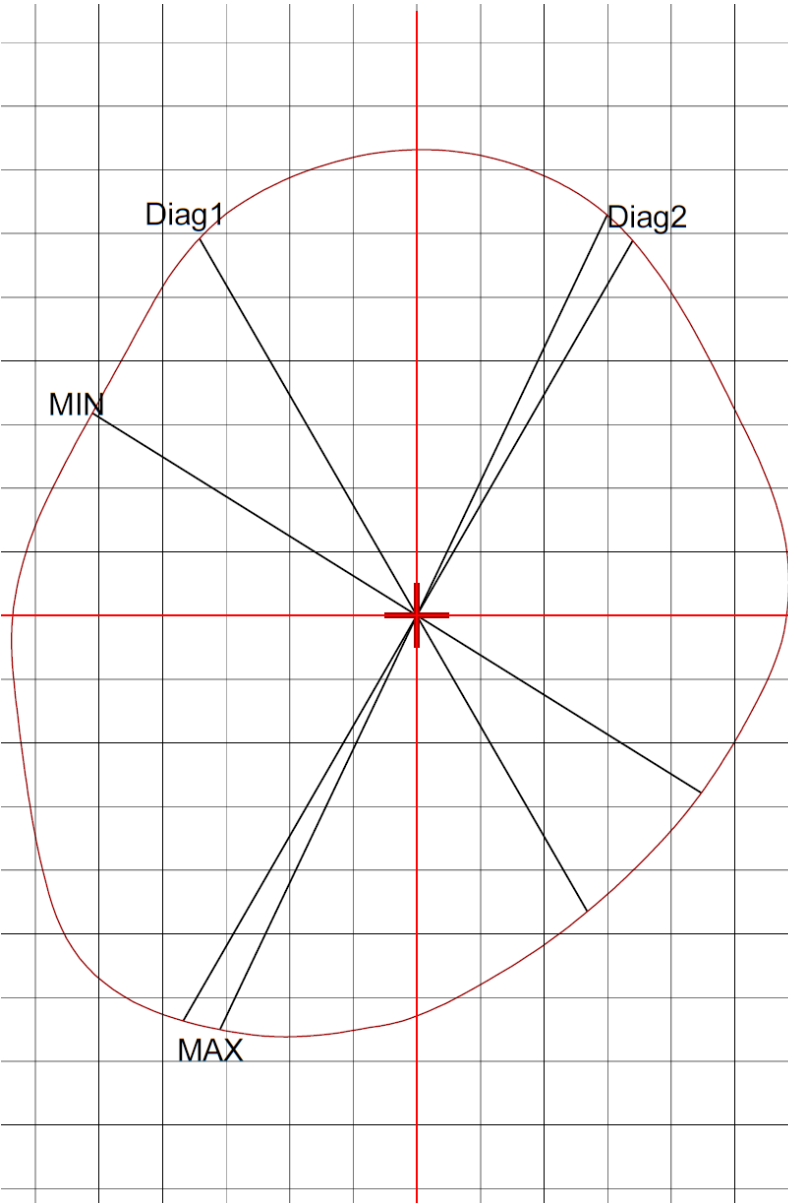


Transdiagonal difference (transcranial diagonal difference)

The difference between two transcranial diameters

Scan





File 1: 110718.dat
 Date: 7/18/2011

Standard Measurements		
Level: 3		
HT:	35.1	
AP:	139.3	
ML:	122.3	
CR:	0.877	
CIRC:	408.5	
RSI:	90.0	
Oblique Measurements		
Diag1:	122.0	at -30.0
Diag2:	141.5	at 30.0
Diff:	19.5	
MIN:	112.8	at -58.1
MAX:	141.7	at 25.5
Selected Measurements		
Dist:		
D1		
D2		

P/R

Cognitive Outcomes and Positional Plagiocephaly

Pediatrics. 2019;

Plagiocephaly and Developmental Delay: A Systematic Review

(J Dev Behav Pediatr 38:67–78, 2017)

Phase 1 (Age < 42 Months)

Controls
N = 237

Children with PPB
N = 235

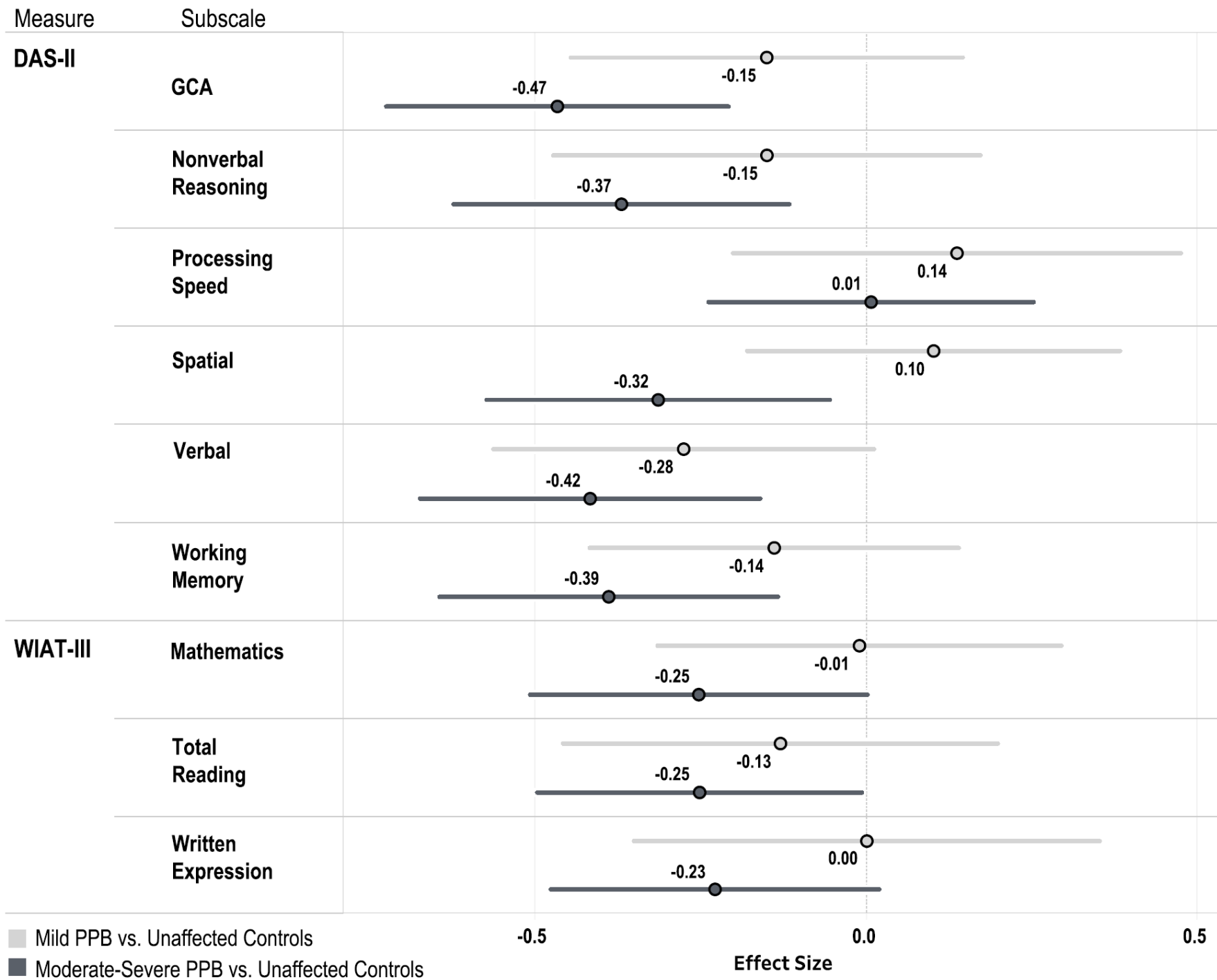
Excluded
PPB (*n* = 70)
Other conditions (Chiari malformation, neurofibromatosis) (*n* = 2)
Child deceased (*n* = 1)

Excluded
No PPB (*n* = 2)
Other conditions (mitochondrial disease, Chiari malformation, genetic anomalies) (*n* = 7)
Child deceased (*n* = 1)

Phase 2 (~8 Years)

Eligible Controls
N = 164

Eligible children with PPB
N = 225
(Mild *n* = 56, moderate to severe *n* = 169)



In summary

- Positional plagiocephaly (brachycephaly)
 - Risk factors: neck ROM, up-right time, sleeping position
 - Naturally regression in most cases
 - Mostly benign cosmetic condition
 - A marker of delayed development in severe cases

Management

- Re-positioning
- Physical therapy
- Orthosis

Helmet therapy

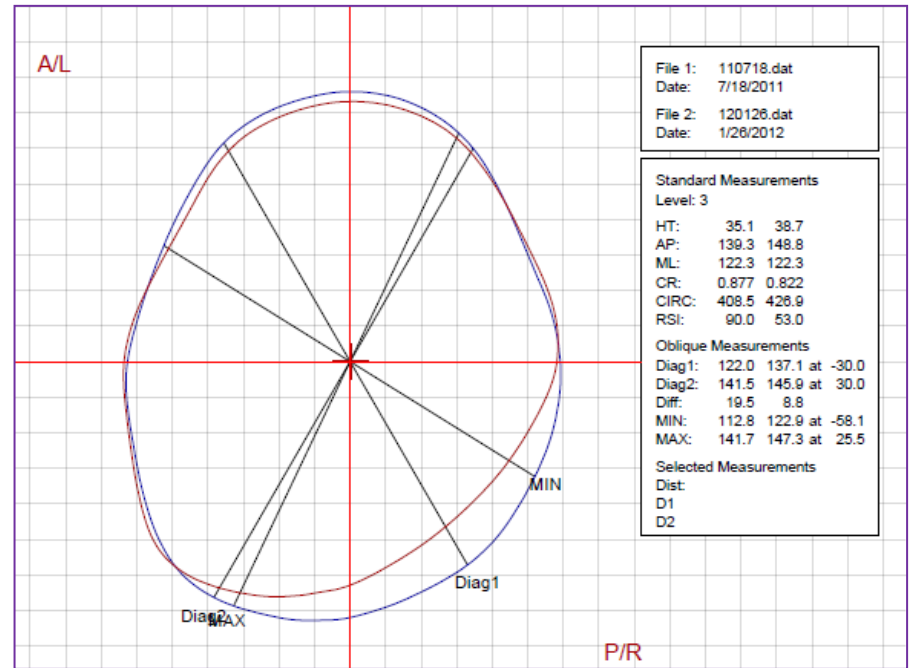




View inside helmet from above



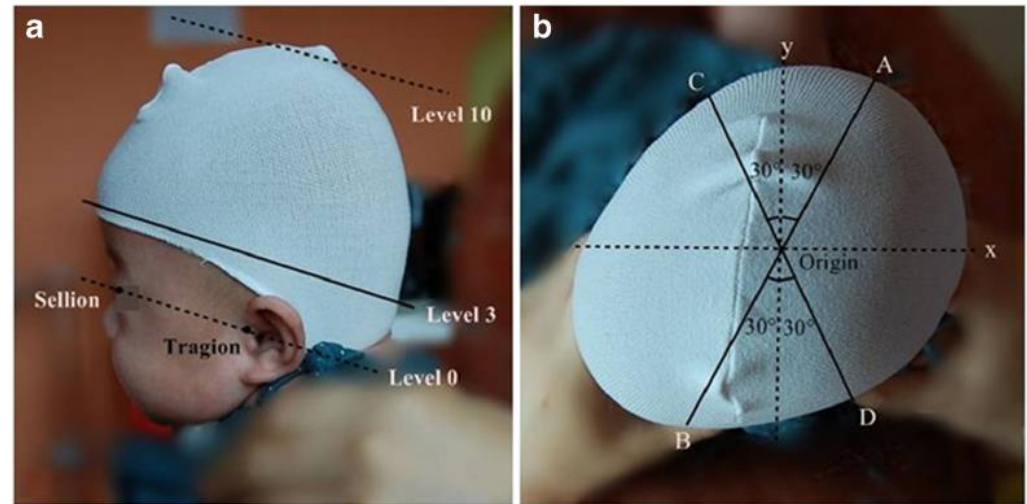
© AboutKidsHealth.ca



Relationship between starting age of cranial-remolding-orthosis therapy and effectiveness of treatment in children with deformational plagiocephaly

Mi-hyang Han¹ • Jin Young Kang² • Hye Young Han¹ • Yun-hwa Cho³ • Dae-Hyun Jang¹

- 310 patients with deformational plagiocephaly
- Cranial vault asymmetry index
$$= \left[\frac{AB - CD}{CD} \right] \times 100$$

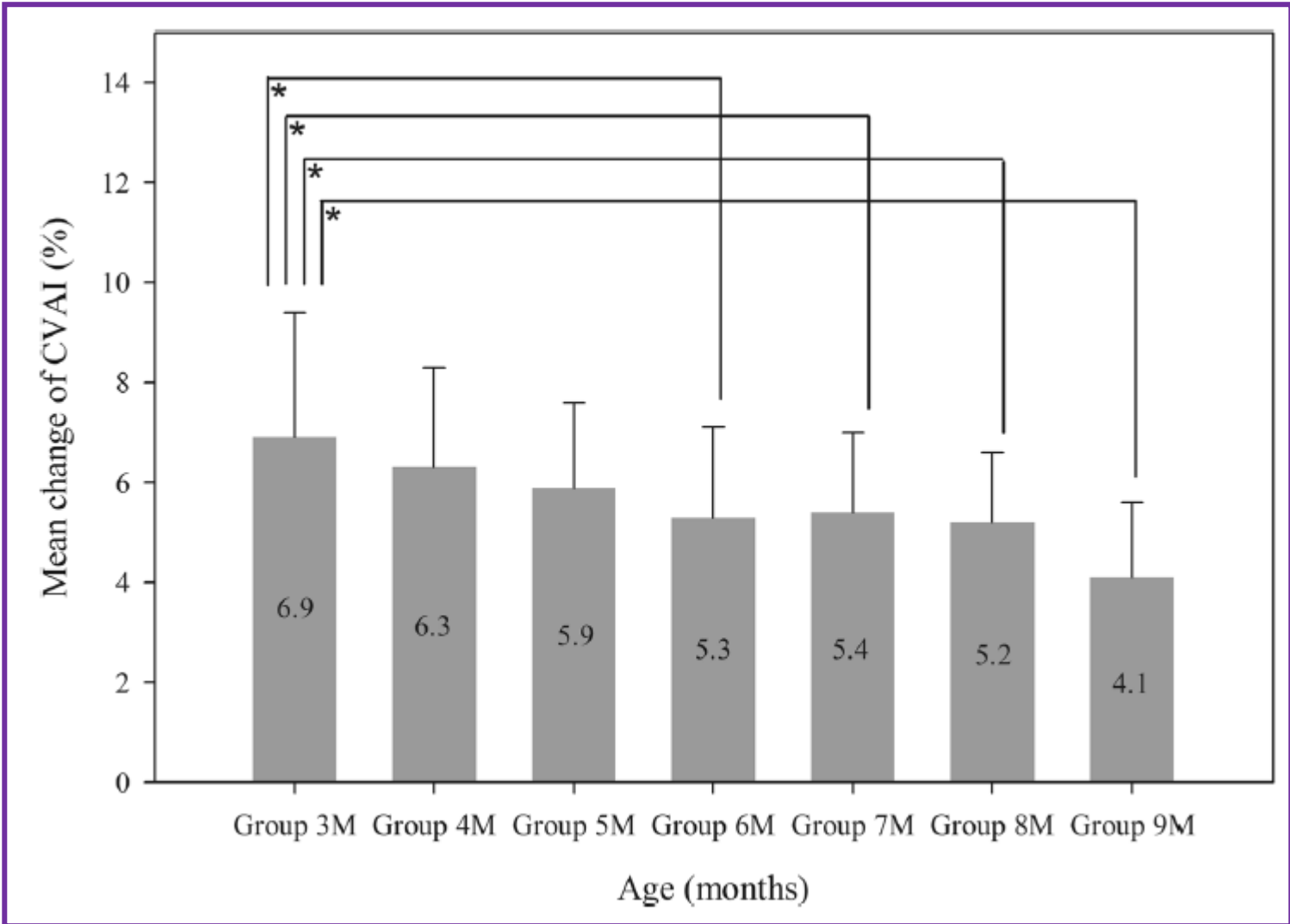


	Total patients	Sex (male/female)	Mean starting age (days)	Severity ^a (mild/moderate/severe)
Group 3M	42	29:13	107.4 ± 11.9 (95.5~119.3)	9:21:12
Group 4M	64	43:21	137.2 ± 8.8 (146~128.4)	6:40:18
Group 5M	66	38:28	162.0 ± 7.8 (154.2~169.8)	9:46:11
Group 6M	56	32:24	194.9 ± 8.4 (186.5~203.3)	11:39:6
Group 7M	32	21:11	224.1 ± 8.1 (216~232.2)	5:23:4
Group 8M	21	12:9	255.4 ± 7.1 (248.3~262.5)	2:15:4
Group 9M	29	12:17	334.0 ± 51.9 (282.1~385.9)	5:19:5
Total	310	187:123	184.3 ± 65.7 (118.6~250.0)	47:203:60

Group 3M, <120 days; group 4M, 120~149 days; group 5M, 150~179 days; group 6M, 180~209 days; group 7M, 210~239 days; group 8M, 240~269 days; group 9M, ≥270 days

^a The initial severity of plagiocephaly was categorized by CVAI as mild group (range, 3.5–7%), moderate group (range, 7–12%), and severe group (range, >12%)



	Duration of therapy (days)	Initial mean CVAI (%)	Final mean CVAI (%)	Mean change of CVAI (%)
Group 3M	124.4 ± 46.9* (77.5~171.3)	10.4 ± 4.1 (6.3~14.5)	3.5 ± 2.1 (1.4~5.6)	6.9 ± 2.5 [‡] (1.4~9.4)
Group 4M	148.0 ± 51.6** (96.4~199.6)	10.1 ± 2.6 (7.5~12.7)	3.8 ± 1.6 (2.2~5.4)	6.3 ± 2.0 ^{‡‡} (4.3~8.3)
Group 5M	156.0 ± 59.6** (96.4~215.6)	9.4 ± 2.4 (7.0~11.8)	3.5 ± 1.5 (2.0~5.0)	5.9 ± 1.7 ^{‡‡‡} (4.2~7.6)
Group 6M	183.4 ± 81.9 (101.5~265.3)	8.8 ± 2.4 (6.4~11.2)	3.5 ± 1.4 (2.1~4.9)	5.3 ± 1.8 (3.5~7.1)
Group 7M	190.8 ± 60.3 (130.5~251.1)	9.9 ± 2.7 (7.2~12.6)	4.5 ± 2.1 (2.4~6.6)	5.4 ± 1.6 (3.8~7.0)
Group 8M	222.0 ± 68.1 (153.9~290.1)	10.0 ± 2.1 (7.9~12.1)	4.8 ± 1.0 (3.8~5.8)	5.2 ± 1.4 (3.8~6.6)
Group 9M	179.5 ± 51.0 (128.5~230.5)	9.8 ± 2.5 (7.3~12.3)	5.7 ± 2.6 (3.1~8.3)	4.1 ± 1.5 (2.6~5.6)
Total	165.3 ± 65.9 (99.4~231.2)	9.7 ± 2.8 (6.9~12.5)	4.0 ± 1.9 (2.1~5.9)	5.7 ± 2.0 (3.7~7.7)



- Craniosynostosis

- Non-synostotic plagiocephaly

Synostotic vs Deformational (Positional)

Characteristics	Positional plagiocephaly	Lambdoid craniosynostosis
Age at onset	Several weeks postnatally	Birth
Preferred position	Common	Rare
Torticollis	Present	Absent/Present
Bony ridge along the lambdoid suture	Absent	Present
Bulging mastoid	Absent	Present
Frontal bossing	Ipsilateral	Contralateral
Displacement of the ipsilateral ear	Anterior	Posterior
Skull shape	Parallelogram 	Trapezoid 
Diagnosis	Clinical, through medical history and physical examination	Three-dimensional computed tomography
Treatment	Clinical	Surgical

3D, three-dimensional; CT, computed tomography.

Synostotic

Deformational

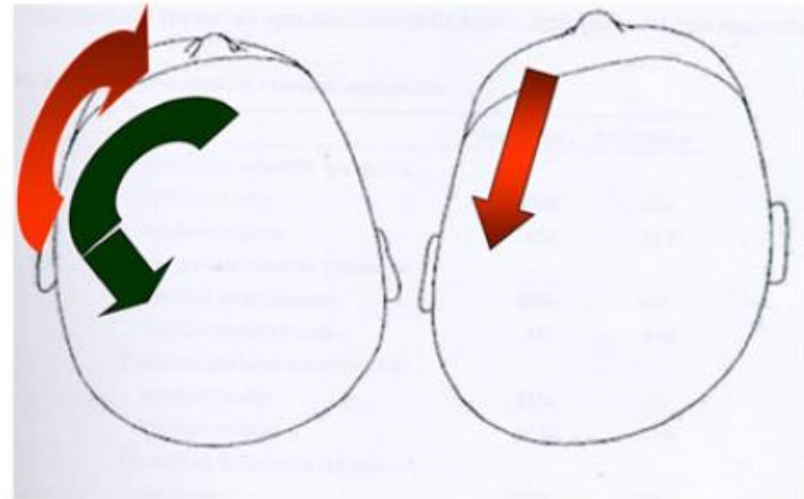
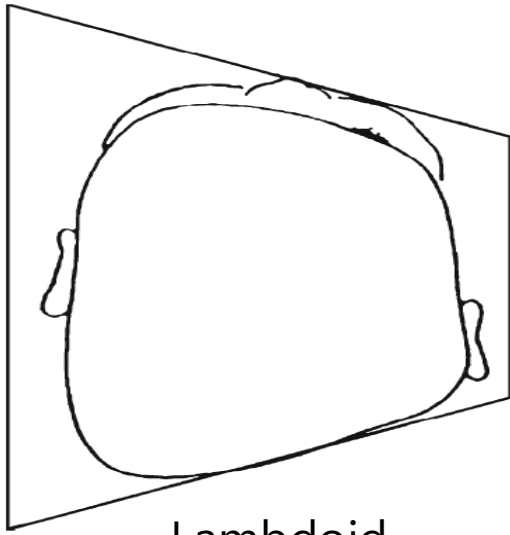
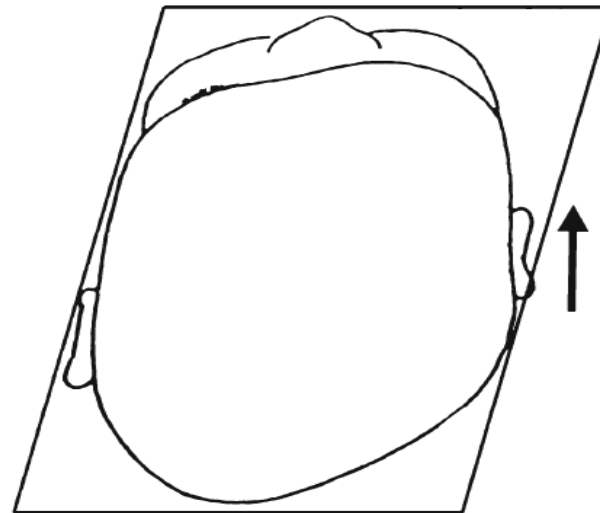


Fig. 3 Dynamic unilateral bone displacement characterizing synostotic (*left*) and deformational (*right*) plagiocephaly

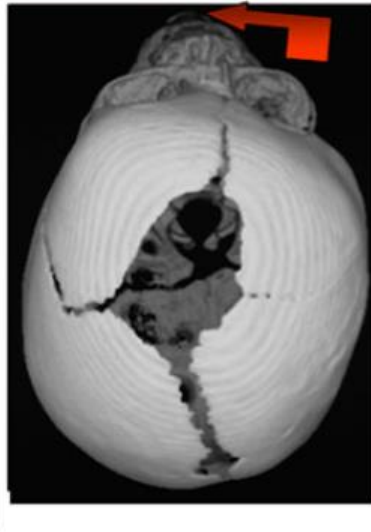
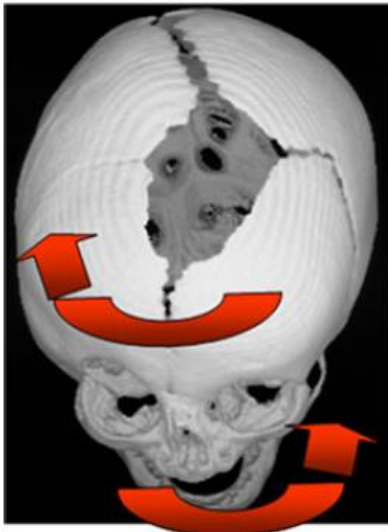
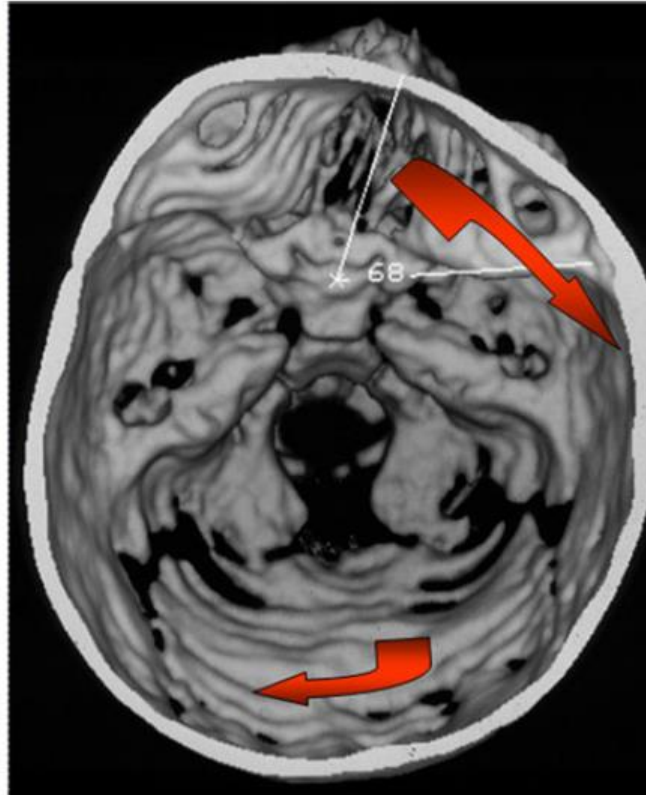
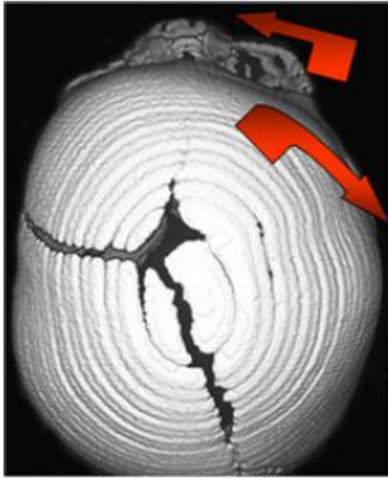


Lambdoid
synostosis



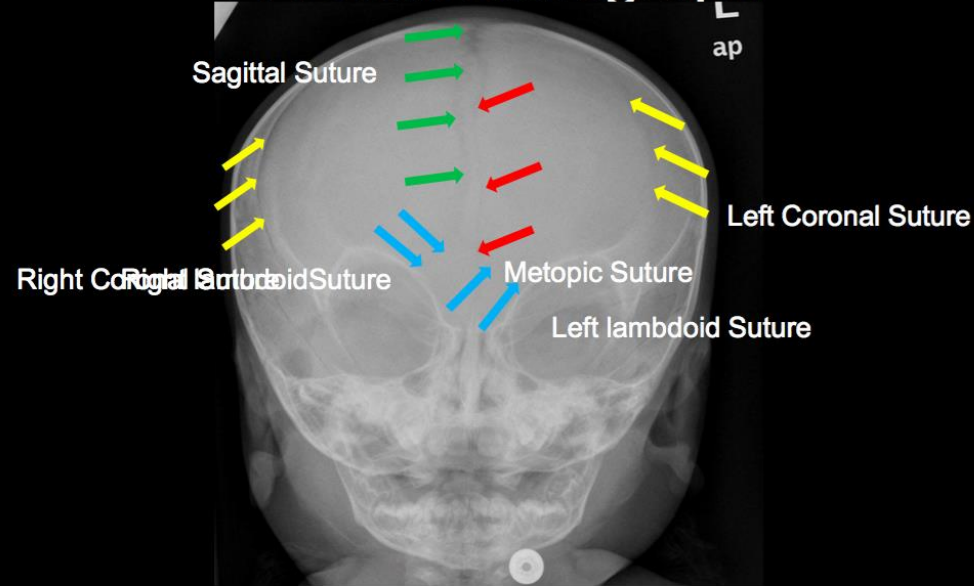
Positional

Craniosynostosis

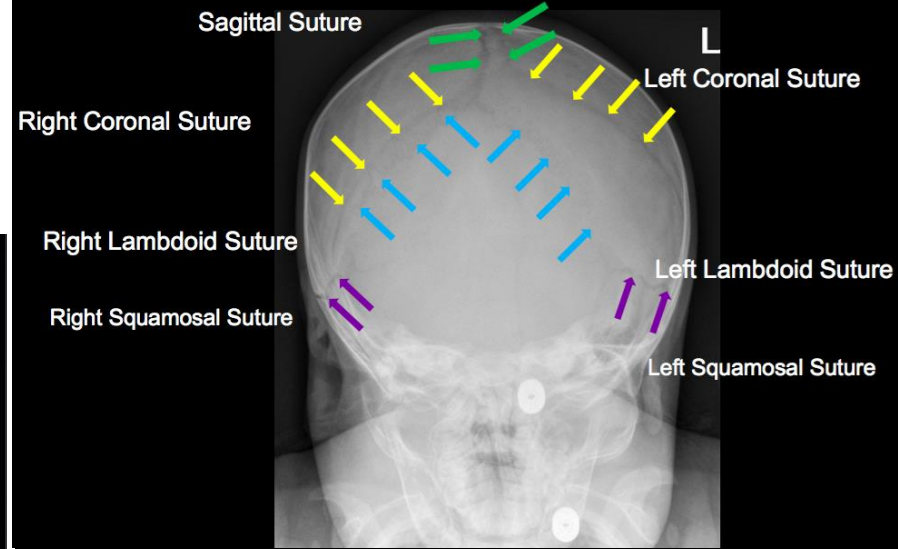


Skull x-ray

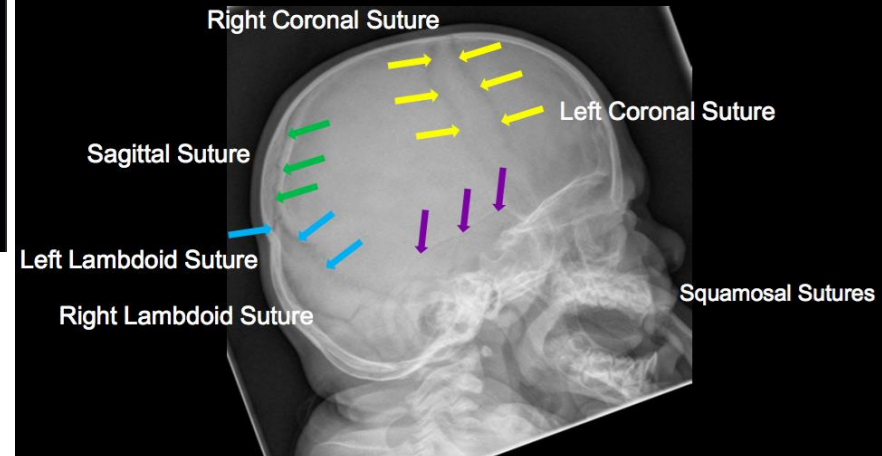
PA Skull radiograph



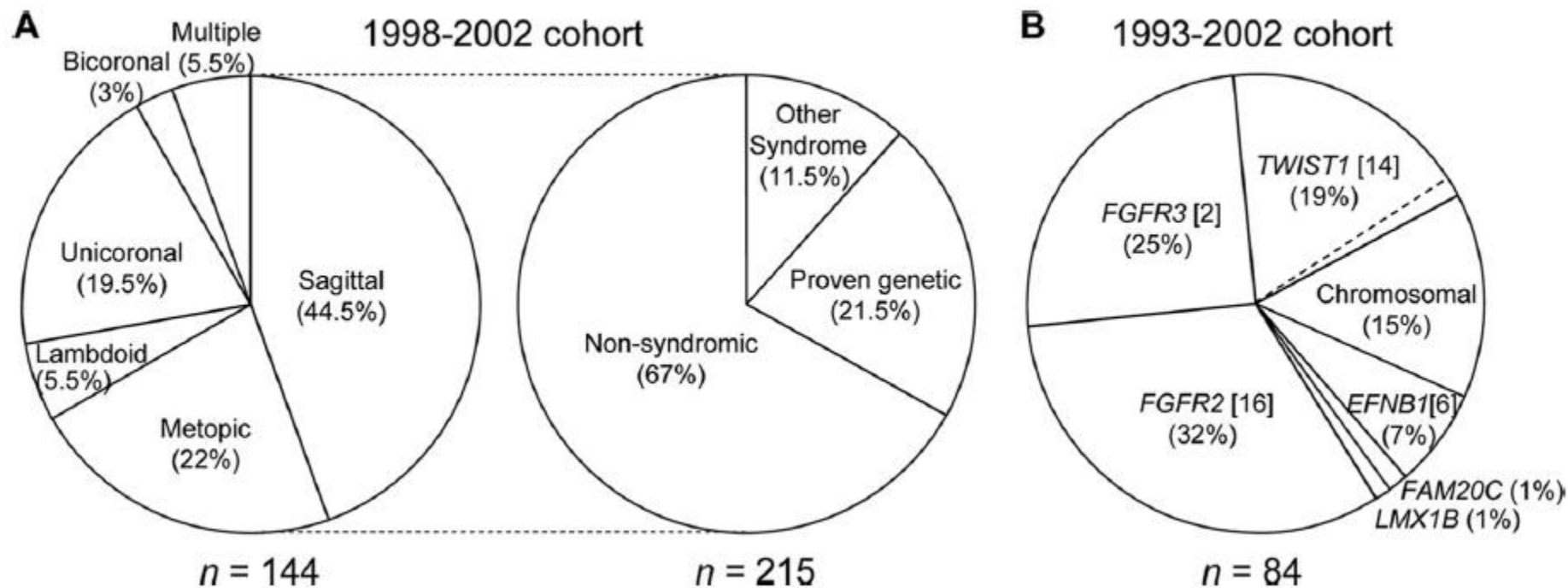
Townes View



Lateral Skull Radiograph



Prevalence and Complications of Single-Gene and Chromosomal Disorders in Craniosynostosis



Increased Incidence of Metopic Suture Abnormalities in Children With Positional Plagiocephaly

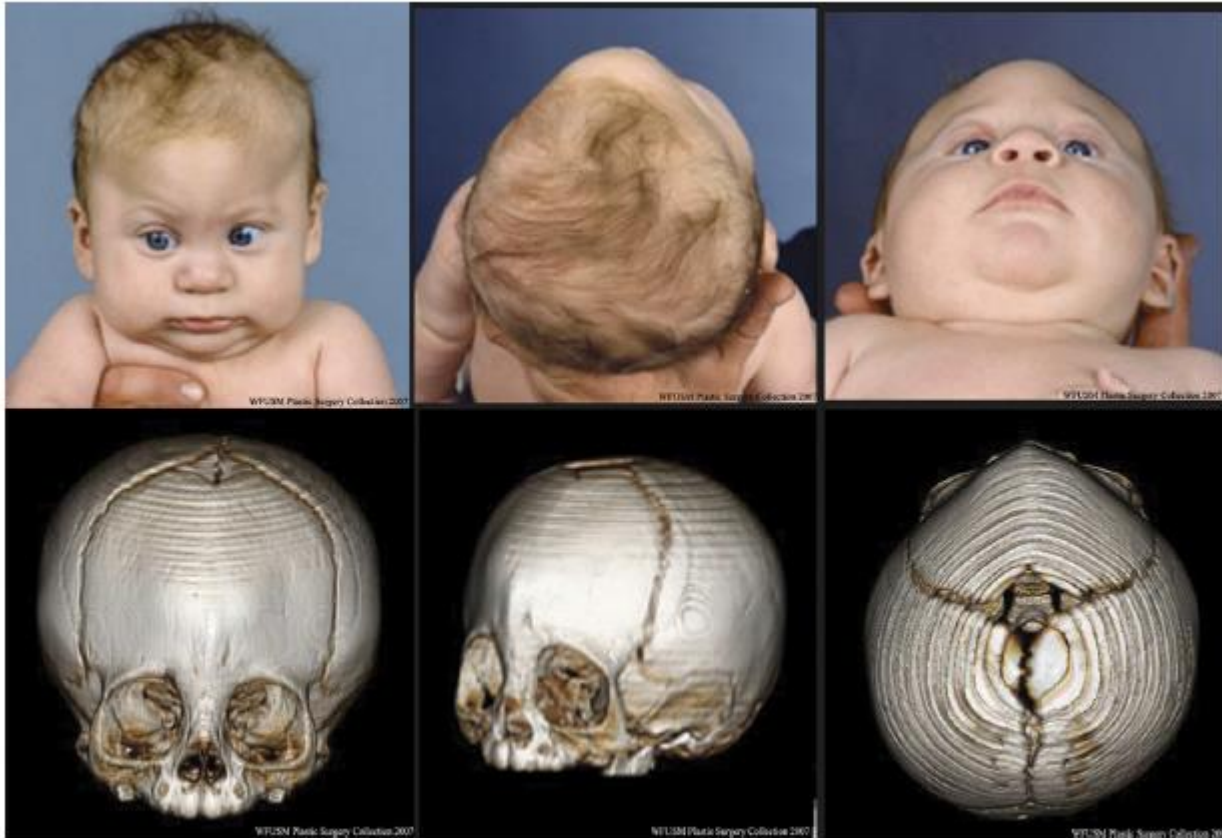


FIGURE 7. Clinical and CT scans of a child with MCS.

(*J Craniofac Surg* 2011;22: 89–95)

Genetics of Craniosynostosis

Genetics of craniosynostosis

- Syndromic
- Nonsyndromic



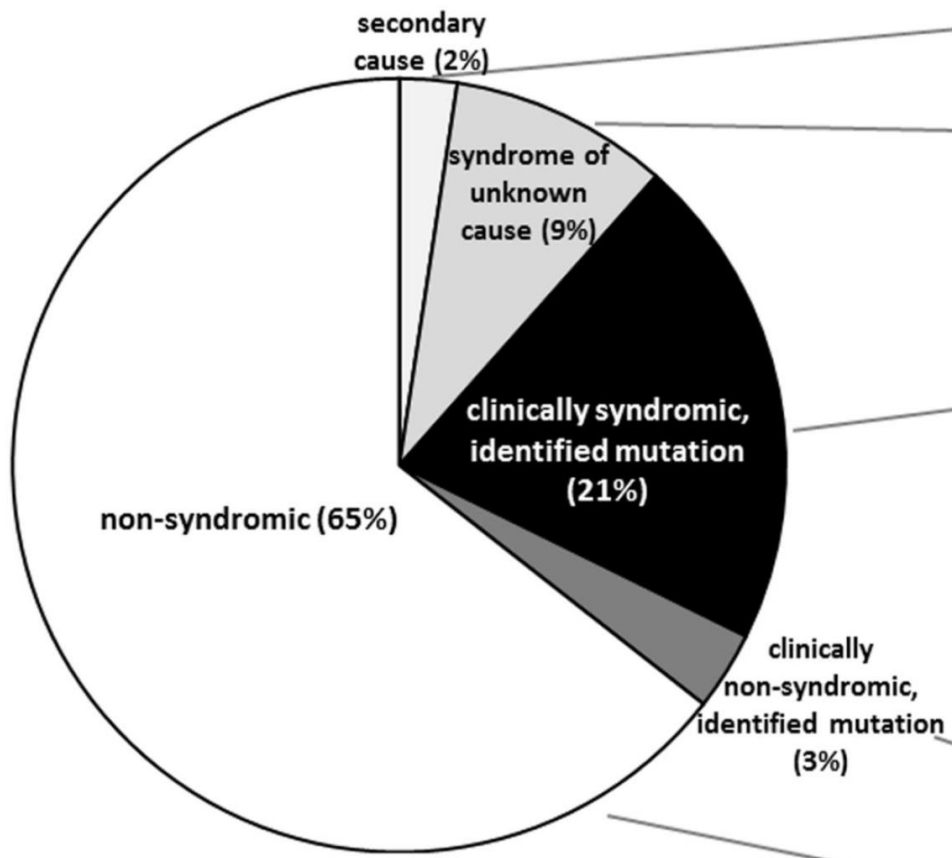
Figure 1: Case 1:(A) Crouzon's syndrome with bilateral proptosis, fusion and elevation of the coronal suture, open anterior fontanelle and brachycephaly. (B and C) "Mitten-type" syndactyly of the fingers and toes with a broad and short thumb are seen

Genetics of craniosynostosis

	Apert syndrome	Crouzon syndrome	Pfeiffer syndrome
Genetic cause	<i>FGFR2</i> : Ser252Trp, Pro253Arg	<i>FGFR2</i> : multiple mutations	<i>FGFR1</i> : Pro252Arg <i>FGFR2</i> : multiple mutations
Skull phenotype	Bilateral coronal synostosis	Bilateral coronal synostosis, pancranio-synostosis, cloverleaf skull	Bilateral coronal synostosis, cloverleaf skull in type 2 Pfeiffer

Muenke syndrome	Saethre–Chotzen syndrome	Craniofrontonasal syndrome
<i>FGFR3</i> : Pro250Arg in all patients	<i>TWIST</i> : multiple mutations, rarely deletions	<i>EFNB1</i> : multiple mutations, rarely deletions; X-linked
Unilateral or bilateral coronal synostosis, macrocephaly	Unilateral or bilateral coronal synostosis, metopic synostosis	Unilateral or bilateral coronal synostosis

666 cohort



suture fused	S	M	UC	BC	L	MS
total	275	129	108	68	9	77
% solved cases	8	13.2	24.1	88.2	11.1	64.9
extreme prematurity	n=7	3	2			2
maternal valproate	n=6		6			
other secondary causes	n=3	1		1		1
syndrome of unknown cause	n=61	22	15	10	4	10
FGFR2-Apert	n=24			19		5
other monogenic	n=23	6	2		2	13
TWIST1-Saethre-Chotzen	n=22			6	12	4
FGFR3-P250R (Muenke)	n=18			3	15	
FGFR2-Crouzon	n=17	5			2	10
chromosomal abnormality	n=16	5	7			4
EFNB1-craniofrontonasal	n=5			3	2	
FGFR2-Pfeiffer	n=4					4
ERF	n=4	1		1		2
TCF12	n=3			1	2	
FGFR3-A391E (Crouzon+AN)	n=2					2
TCF12	n=9			6	2	1
FGFR3-P250R (Muenke)	n=6			2	4	
ERF	n=3	1				1
FGFR2	n=2			1		1
TWIST1	n=2			2		
non-syndromic	n=429	231	97	72	4	8

Figure 1. Classification and causes of craniosynostosis in a prospectively ascertained 13-year cohort

Management

Craniosynostosis

- Intracranial hypertension (ICH)
- Cognitive and developmental disorders
- Poor weight gain
- Visual, hearing, and language disorders
- Psychological problems such as low self-esteem and social isolation

Endoscopic versus open approach in craniosynostosis repair: a systematic review and meta-analysis of perioperative outcomes

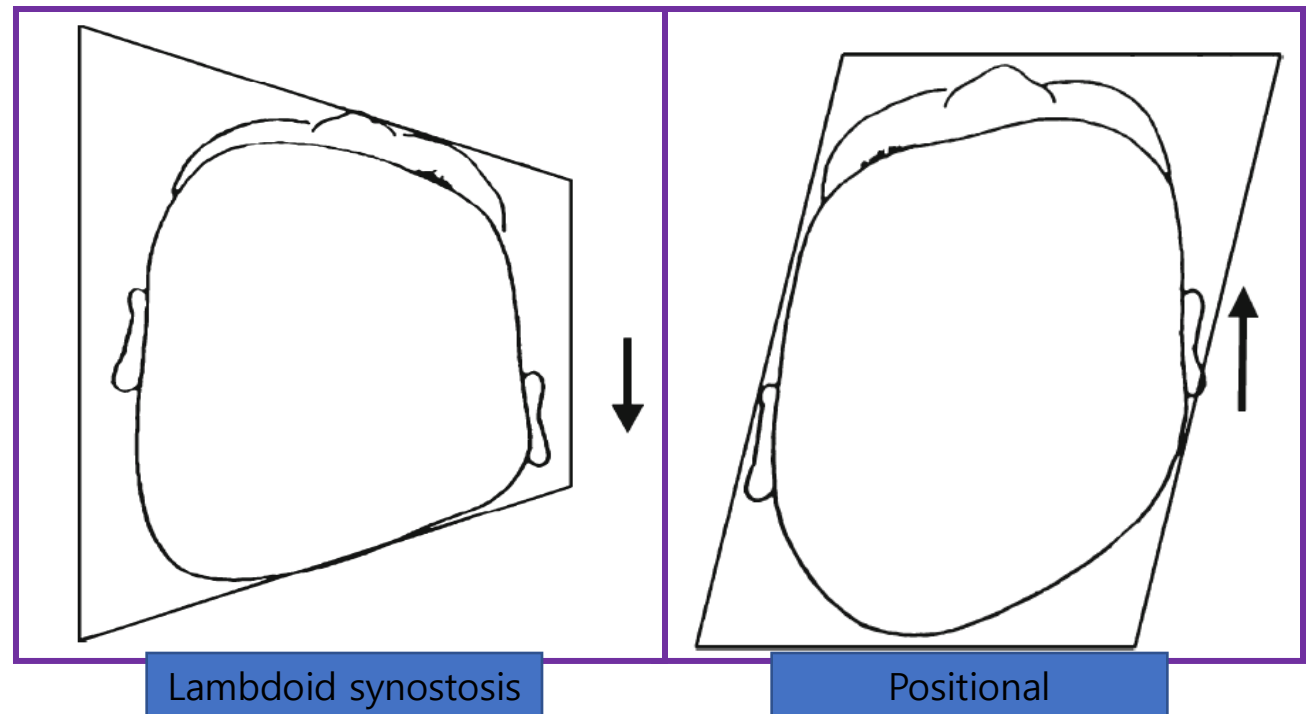
- Blood loss
- Operative time
- Length of stay

Summary

Craniosynostosis

Craniosynostosis

- D/Dx. Deformational plagiocephaly



Craniosynostosis

- Careful P/Ex and skull x-ray
- If necessary, 3D CT
- If possible, genetic studies
- Refer to surgeon as soon as possible

- 감사합니다.