

Trattamento mini invasivo delle malformazioni cranio facciali

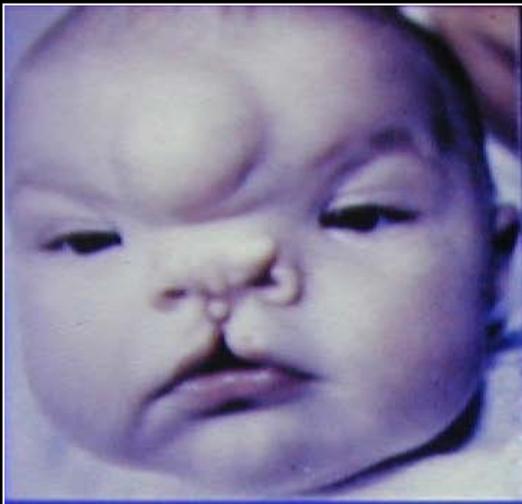
Dalle trasposizioni di lembi alla chirurgia mini-invasiva

Lorenzo Genitori et al.

Divisione di Neurochirurgia
Coordinamento Regionale Neurochirurgia Pediatrica
Regione Toscana
Ospedale Pediatrico “Anna Meyer” - Firenze



Craniofacial surgery especially in children have had a tremendous upgrade in the last 20 years with improvement in surgical skill and standard better results in almost all dedicated center



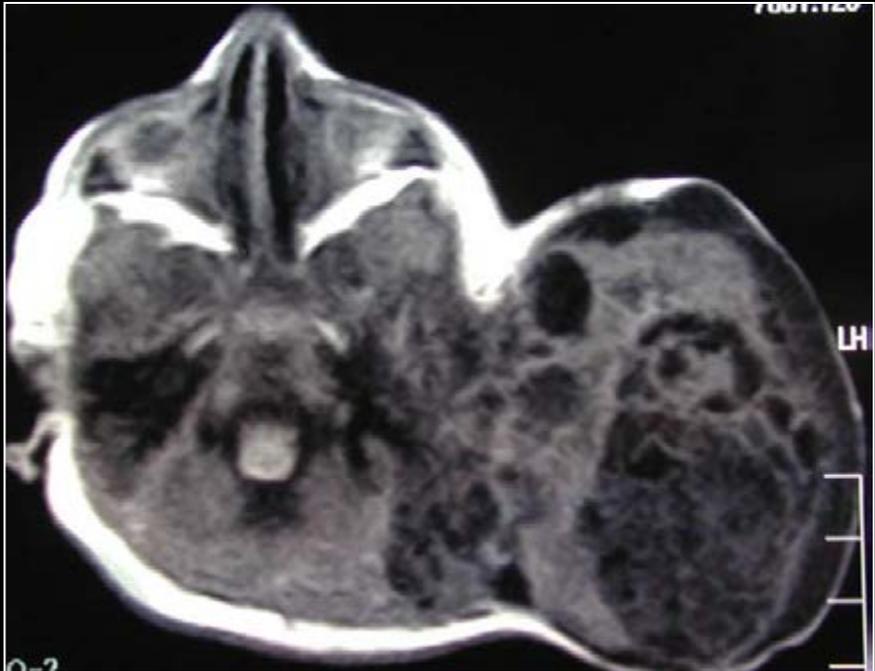
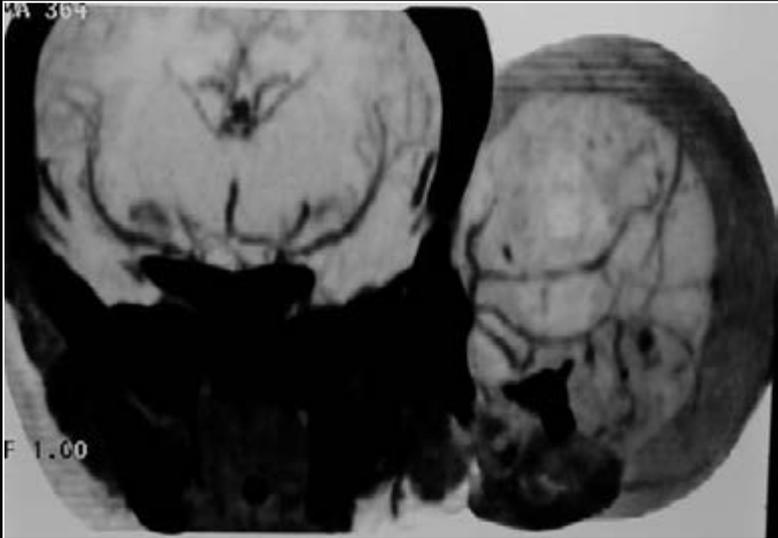
At birth

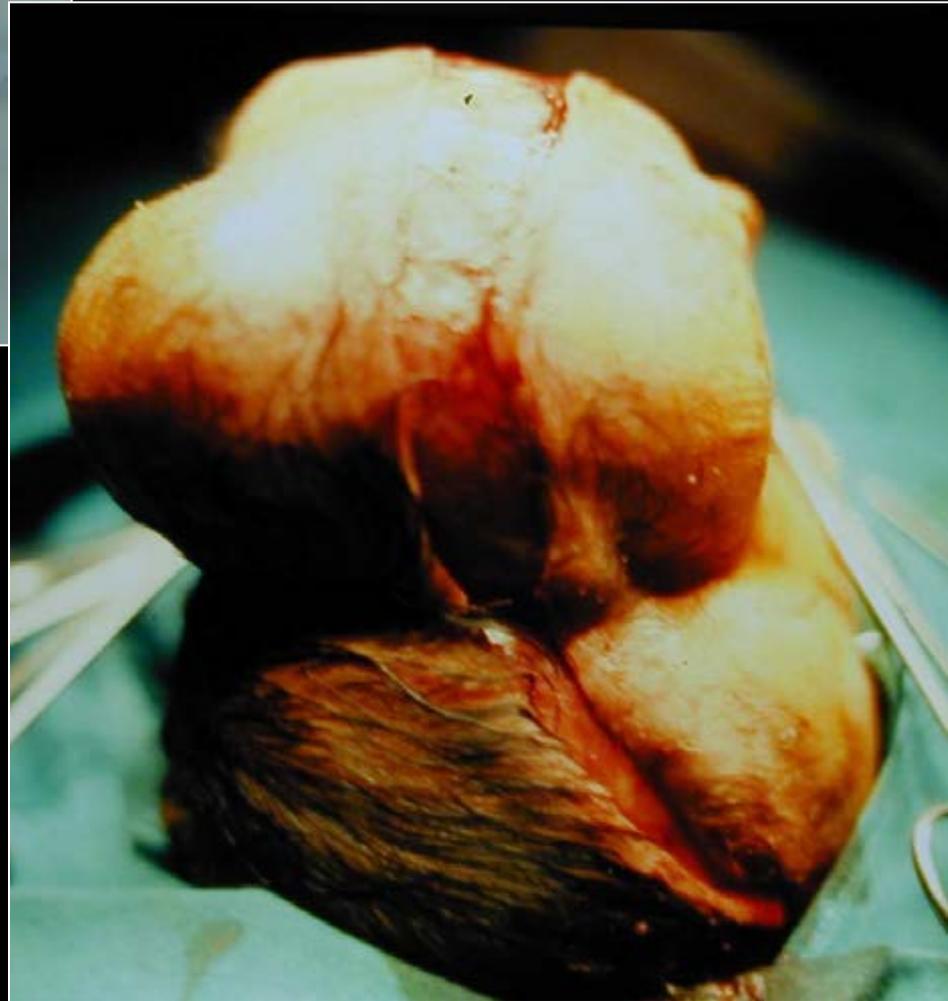


6 years



Giant Teratoma in newborn





.....1 year later



.....1 year later



BUT

- 1) 10% fair or very poor results
- 2) Some patients develop late mental delay
- 3) Some patients will have hydrocephalus
- 4) Asymmetrical forms have worse results





Ingresso e Serra fotovoltaica

Terrazza pensile
con “cappelli di
pinocchio”

Progetto 1999
Trasferimento
dicembre 2007
“Ecosostenibile”





Ospedale organizzato per intensità di cure

171 letti degenza ordinaria

57 letti degenza diurna

7 Dipartimenti

576 personale all'assistenza

52 personale tecnico

194 personale medico



La chirurgia cranio facciale nel bambino e nell'adolescente

2130 casi

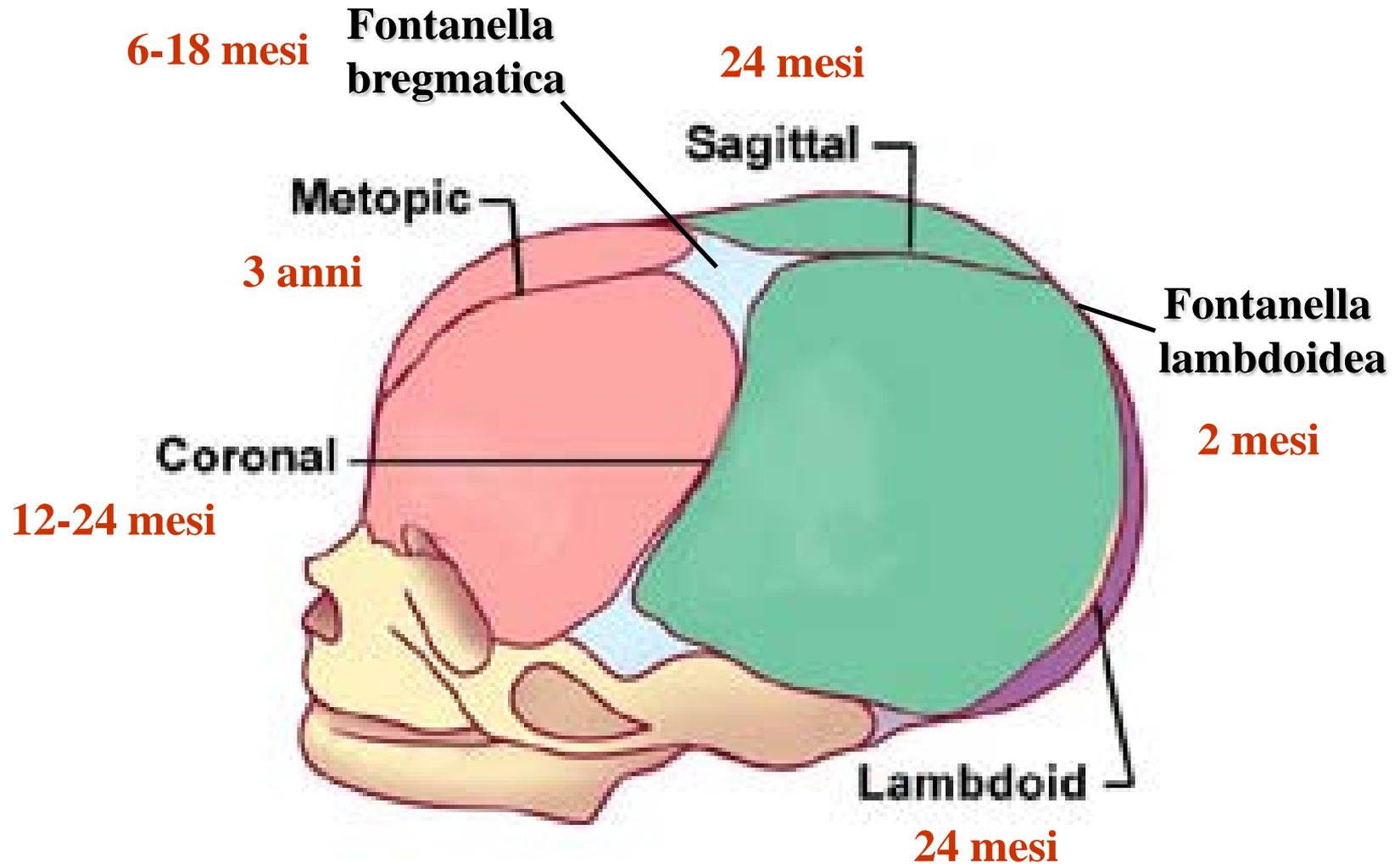
Necessità di
un'approccio
multidisciplinare

Cranio facial surgery team

in children and teen-agers

- . Neurosurgery
- . Orthodontist
- . Genetist
- . Rehabilitation
- . Speech therapy
- . ENT surgery
- . Anesthesiology
- . Maxillo facial
- . Plastic surgery
- . Dentistry
- . Pediatrics
- . Ophtalmology
- . Radiology
- . Psychology

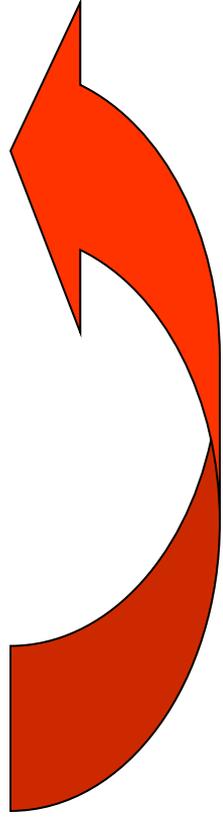
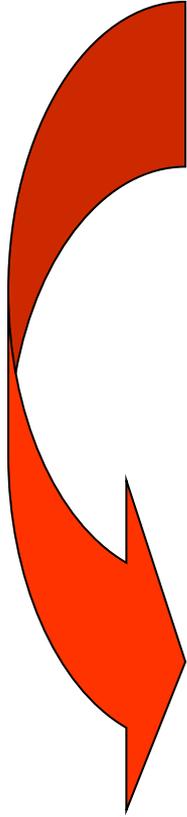
Tempi di chiusura delle suture



Anomalie della volta cranica

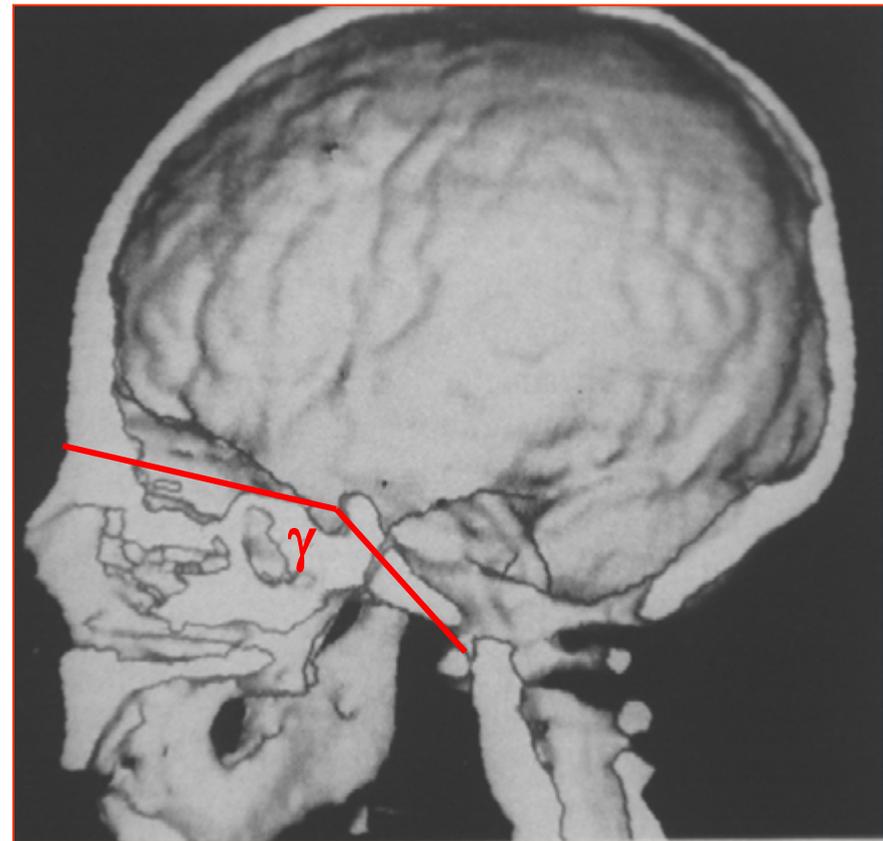
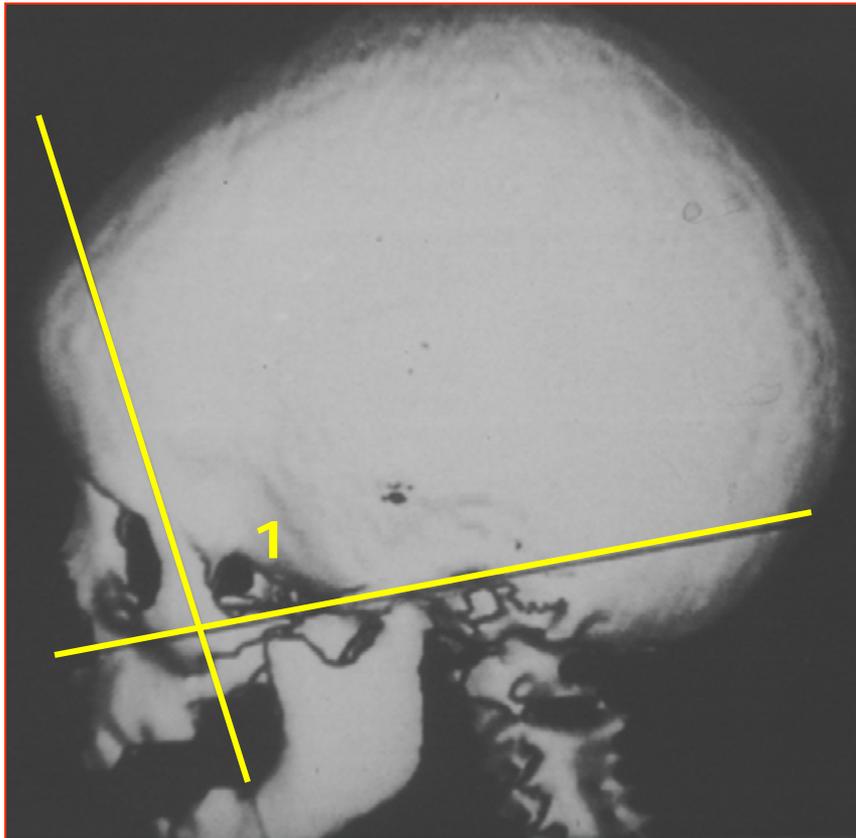
Anomalie strutturali basicranio

Malposizione orbitaria



Valutazione TC – TC 3D

- Orientamento spaziale di zigomo e angolo zigomatico (1)
- Angolo basale di Bogaert (nasion-clino-basion) (130-140°) (γ)



- Angolo nasion-pterion (α)
- Angolo clino-pterion (β)
- Distanza nasion-clinoide (a)
- Distanza bipterionale (b)
- Angolo nasion-clino-pterion (γ)
- Angolo nasion-clino-rocche (δ)
- Orientamento rocche

- Volume emifossa anteriore
- Volume emifossa temporale
- Volume emifossa posteriore

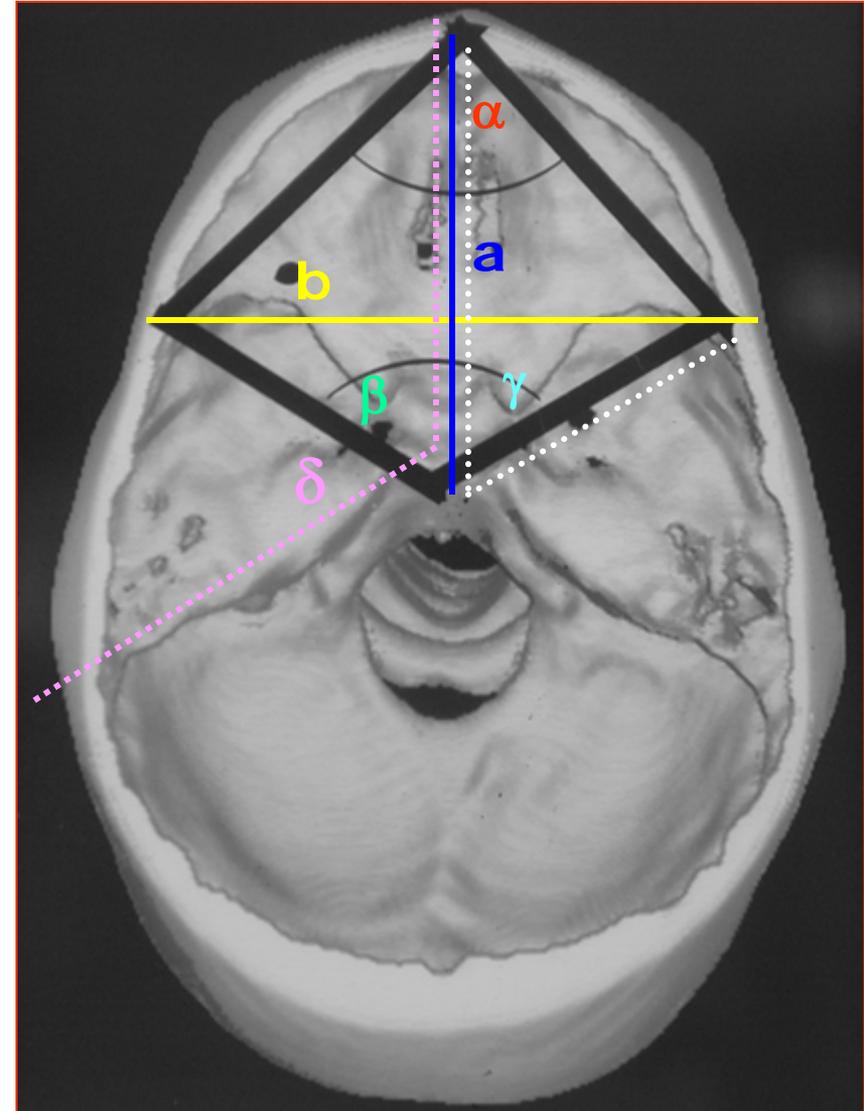
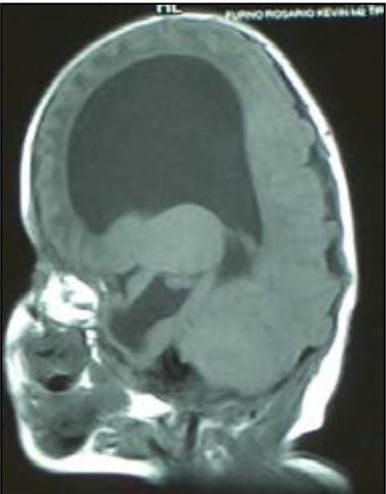
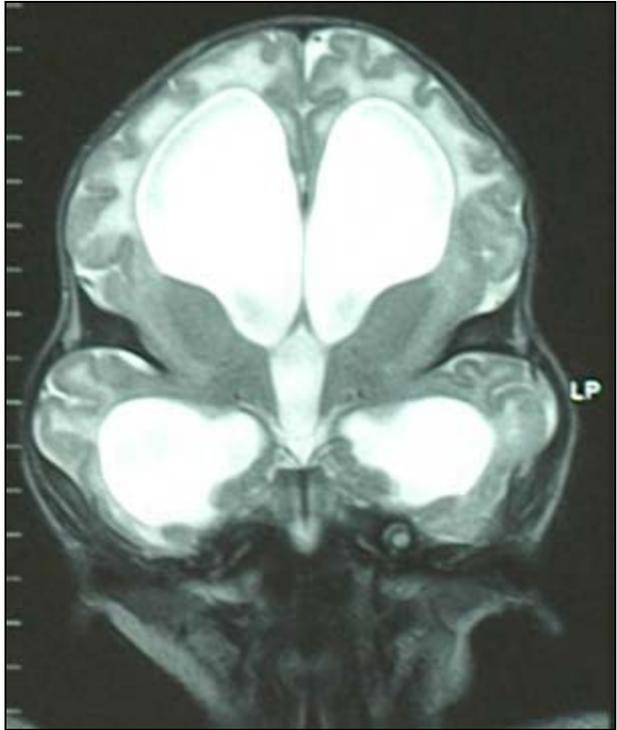
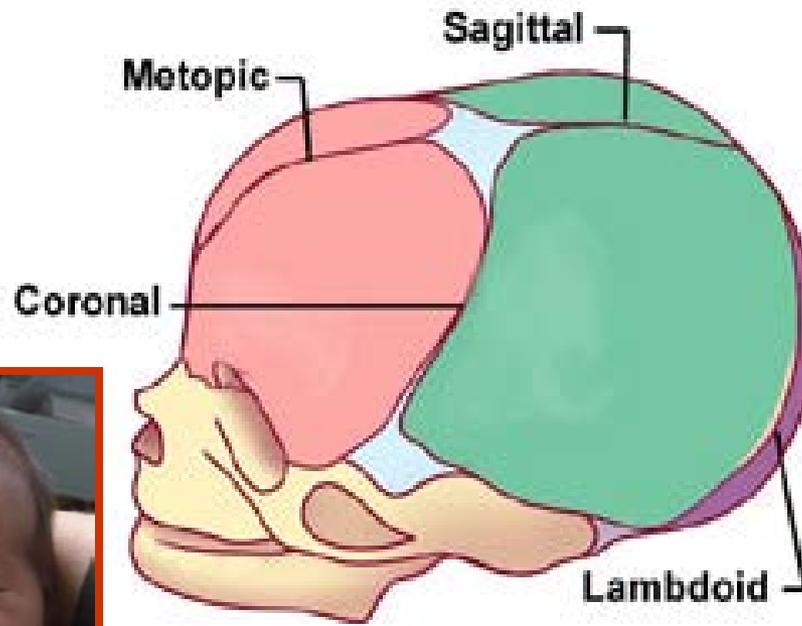




Immagine in Risonanza magnetica





Analisi Genetica (DNA) 1986- 2000

1150 casi trattati: 26 analizzati (2.2%)



Analisi Genetica (DNA) 2000 - 2009

520 casi trattati: 390 analizzati (75%)

Markers Genetici nelle Craniosinostosi

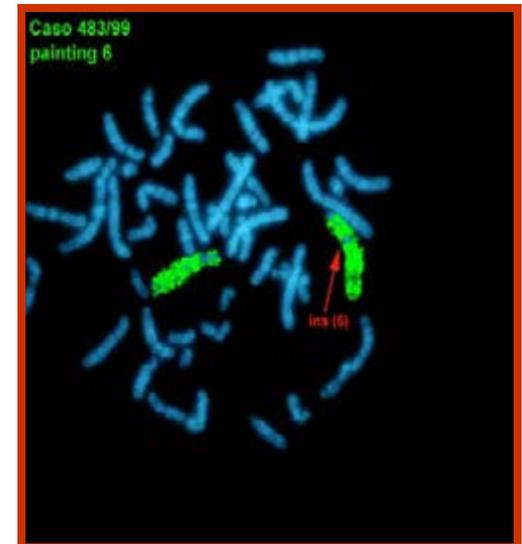
Type	CR	FGR2IIIa (CR 10q)	FGFR2IIIc (CR 10q)	FGFR3 (CR 4p)	TWIST (CR 7p)
Scaphocephaly	-/+	-	-	-	-
Trigonocephaly	-/+	-	-	-	-
Ant Plagiocephaly	-	-	-	+++	+++
Post Plagiocephaly	-	-	-	-	-
Brachicephaly	-	-	-	++	+
Saethre-Chatzen Sdr	-	+	-	++	+++
Crouzon Syndrome	-	++	+++	+	+
Apert Syndrome	-	+++	+	+	+
Pfeiffer Syndrome	-	++	+	+	+

Frequentemente il cariotipo
routinario è normale in morfologia e
numero in questi pazienti

La FISH subtelomerica
ha un ruolo essenziale



Sostituito oggi dall'analisi in Array CGH



Indicazioni e principi di trattamento

- 1) Considerazioni sulla pressione endocranica
- 2) Considerazioni oftalmologiche
- 3) Considerazioni estetiche

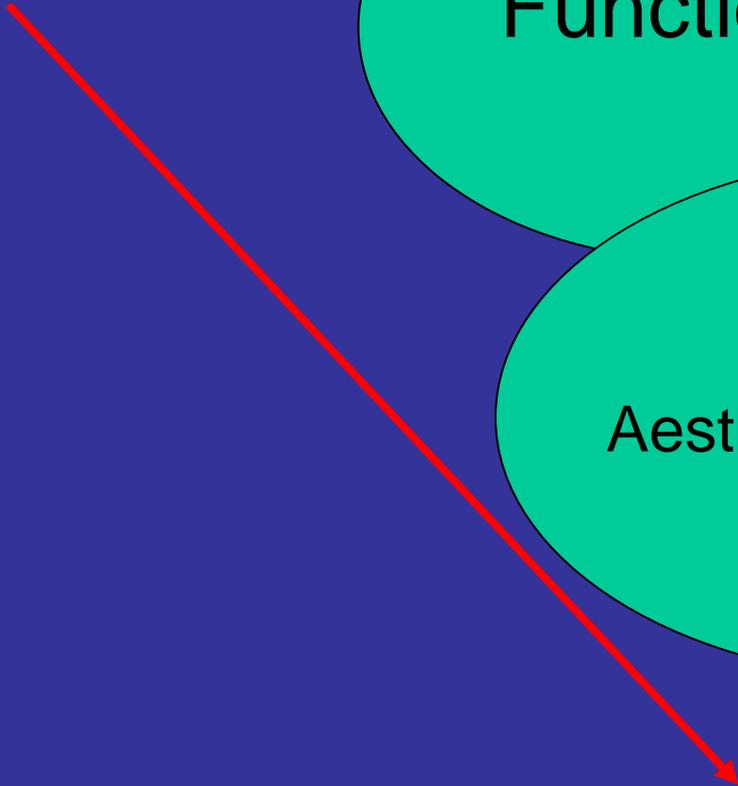
Principles of treatment

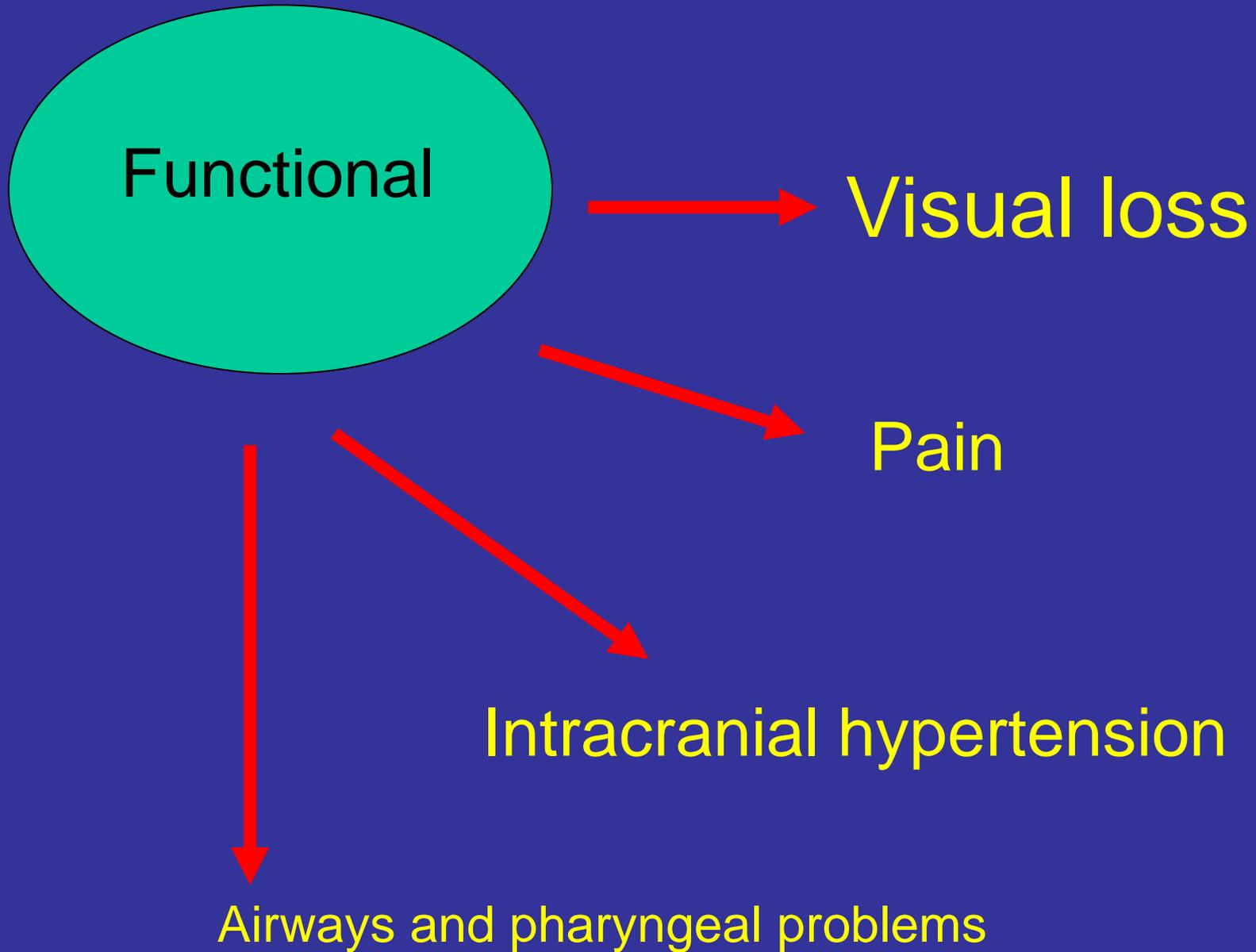
Pediatric age

Functional

Aesthetic considerations

Adult age





Speech problems

Deformity

Proptosis



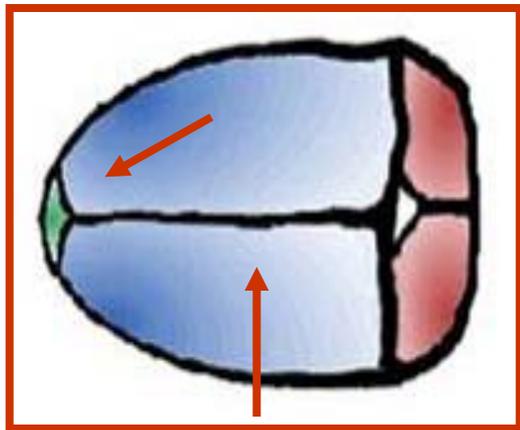
Aesthetic considerations



**Testa lunga e
MACROCRANIA**

Scafocefalia

Chiusura precoce
delle
suture **sagittale** e
talvolta **lambdoidee**



Crescita compensatoria

Aumento del rapporto tra
la distanza
inion-nasion
e la distanza **bitragale**

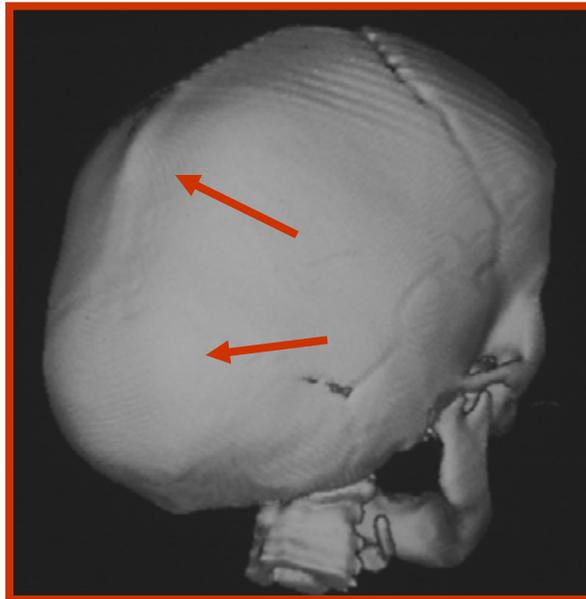


Bozze frontali
prominenti



DIAGNOSI

- **OSSERVAZIONE CLINICA**
- X-ray
- 3D-CT scan
- MRI nel sospetto di malformazioni cerebrali



Scafocefalia

Indicazioni chirurgiche

- Mainly aesthetic
- Controversy about Intracranial Pressure
- NOT surgery in case of "compensated" scaphocephaly (i.e. dolicocephaly)



longitudinal diameter vs transverse diameter \leq 2 cm

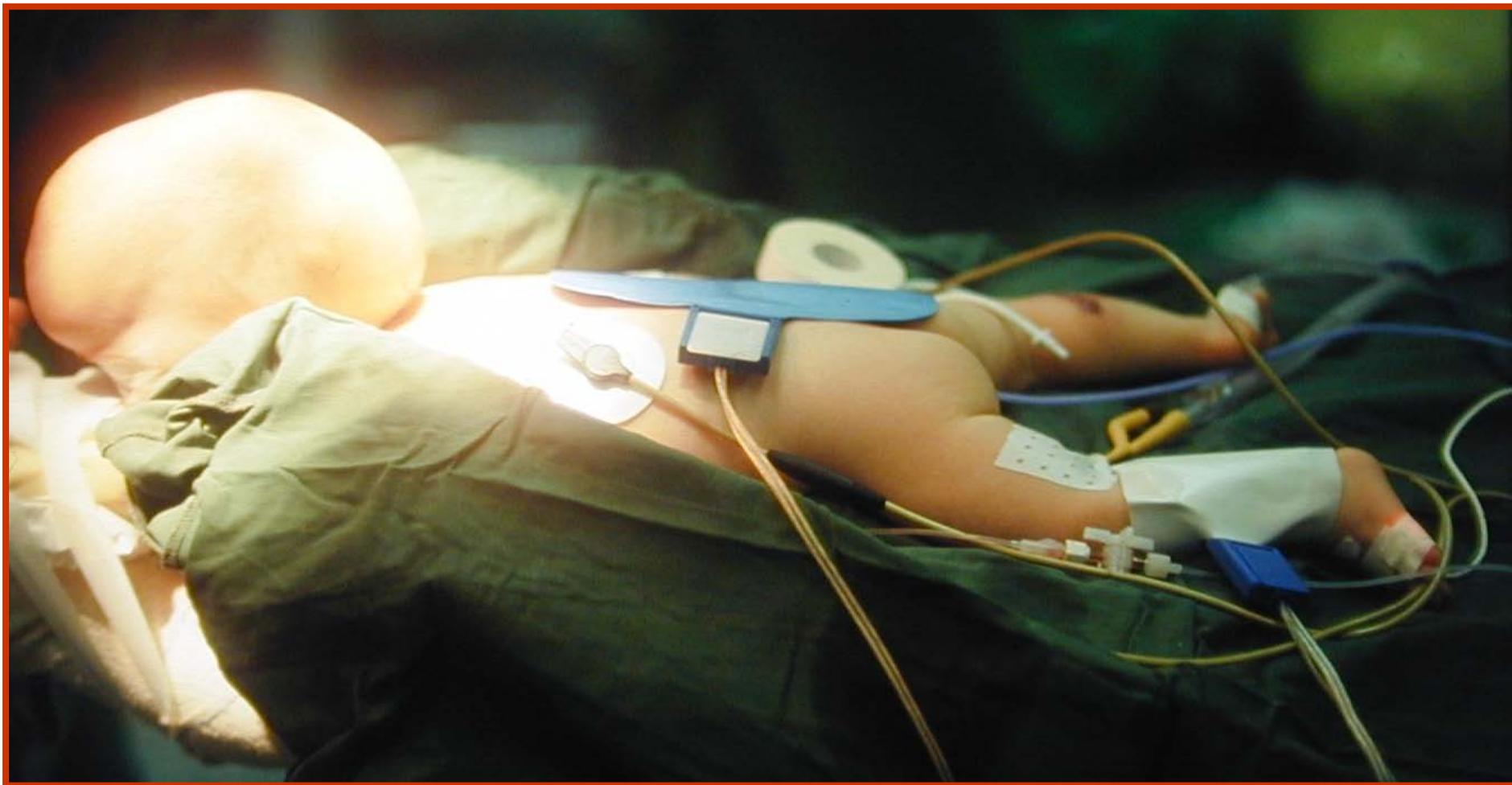
Scafocefalia

Principles of treatment

Ideal age : as soon as possible

- < 6 mth: Linear stripping
of sagittal suture
- > 6 mth: Total cranial vault remodeling
(pi grec, transposition, morcellation)

Posizione sul tavolo operatorio



Incisione cutanea

“S” italice



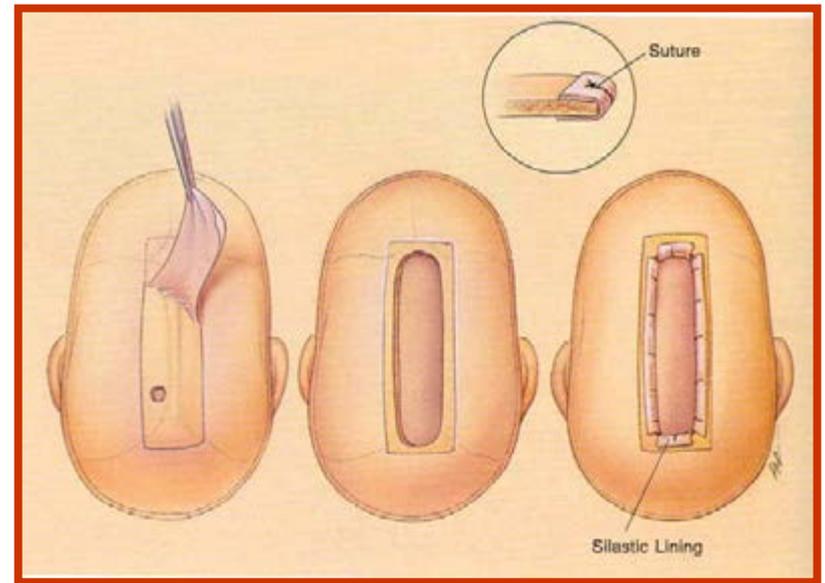
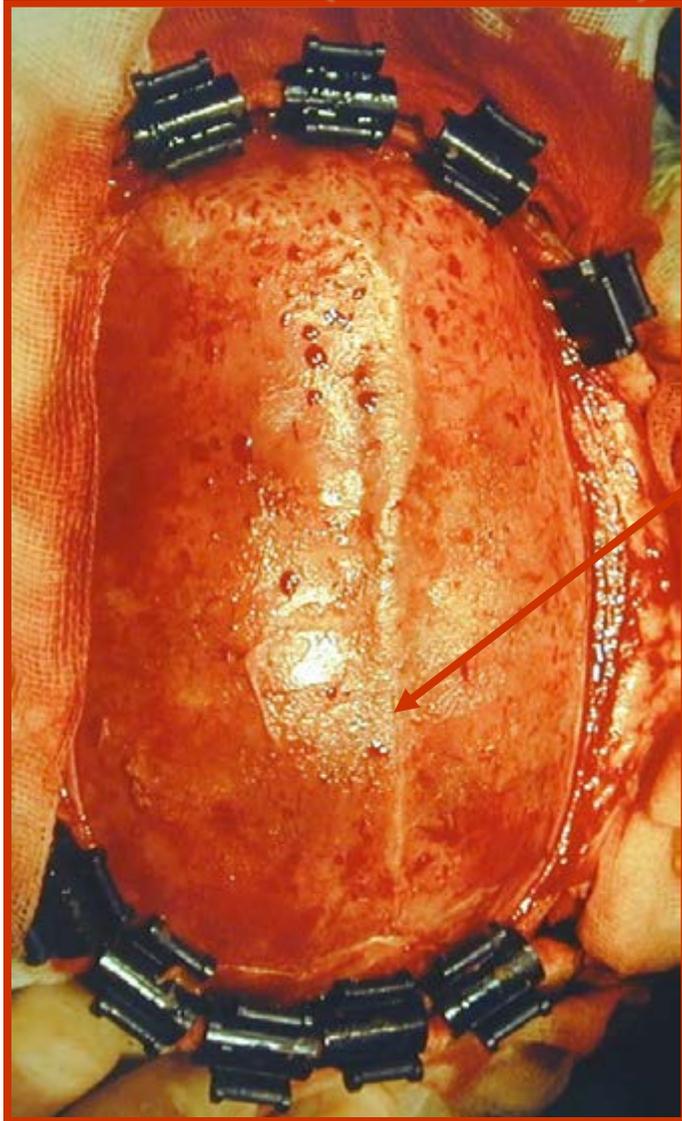
lineare bitragale



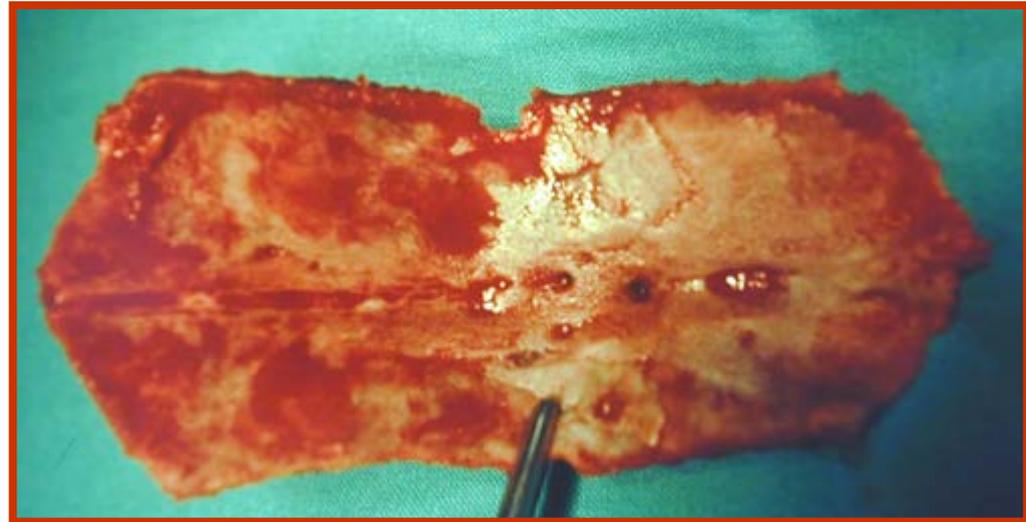
“zig-zag” bitragale

Stripping Sagittale

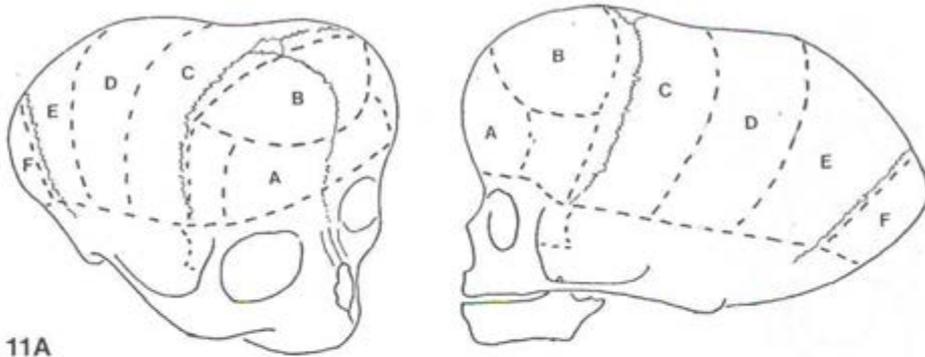
(3-6 mesi)



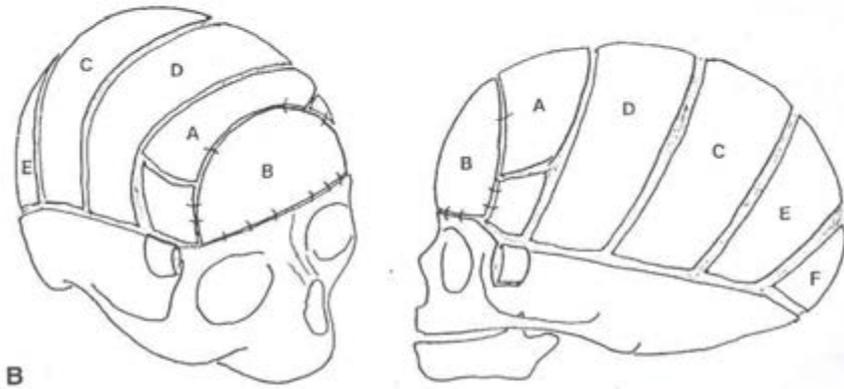
Sutura sagittale



Rimodellamento completo della volta cranica



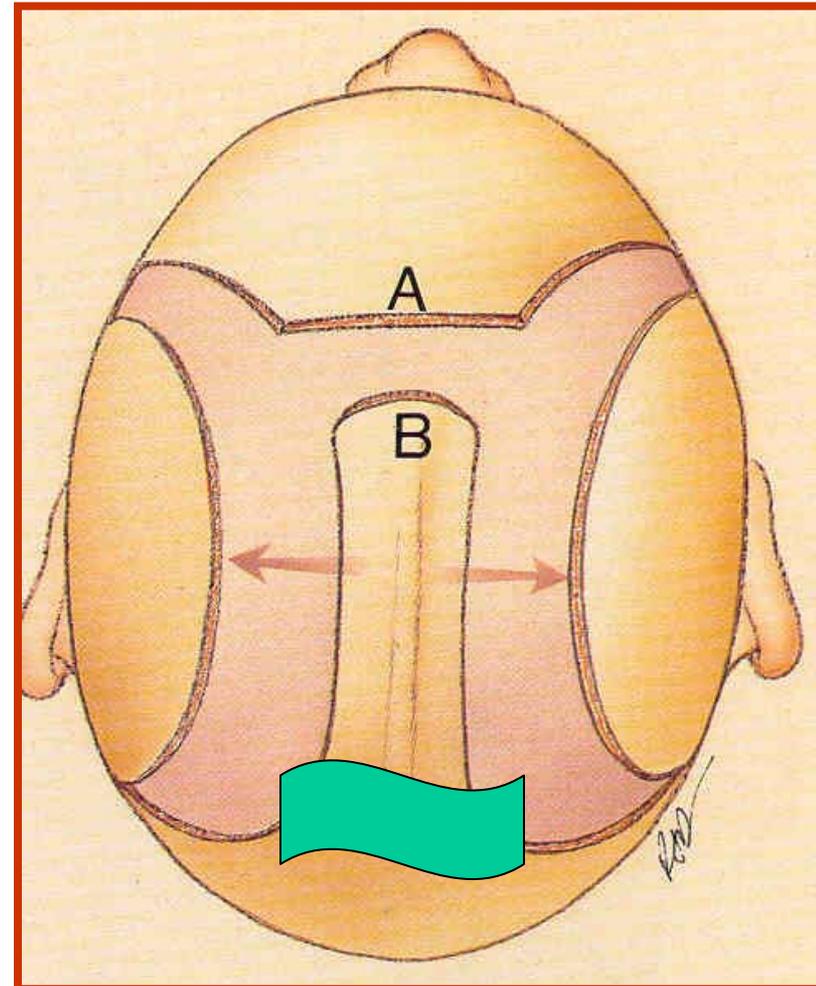
11A

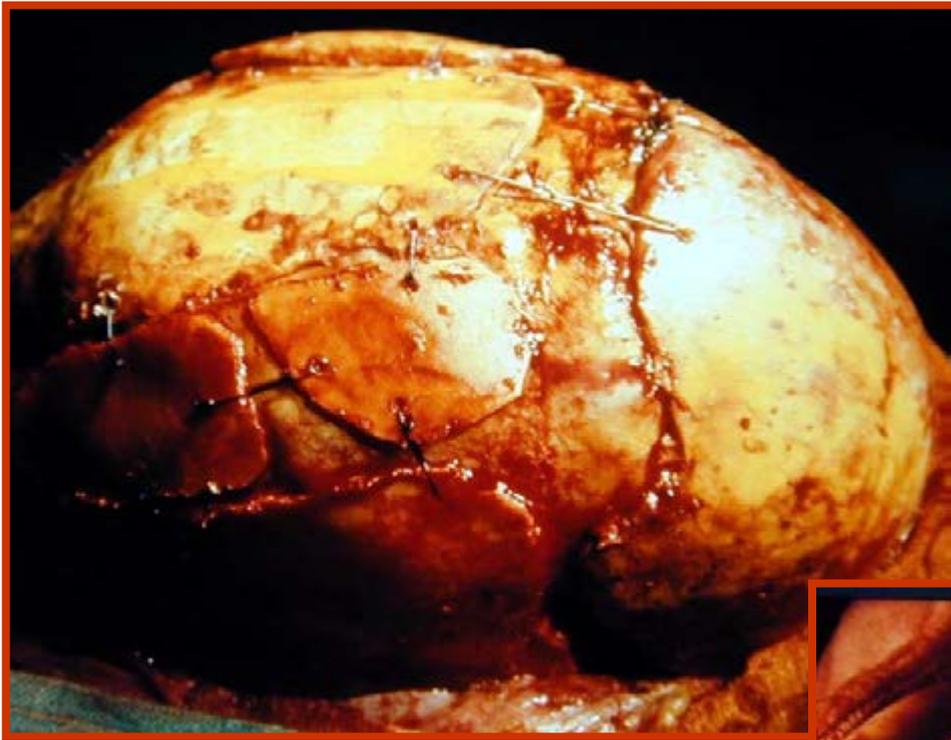


B

DOPO I 6 MESI

“ π ” tecnica





**Complete remodelling
of the cranial vault**

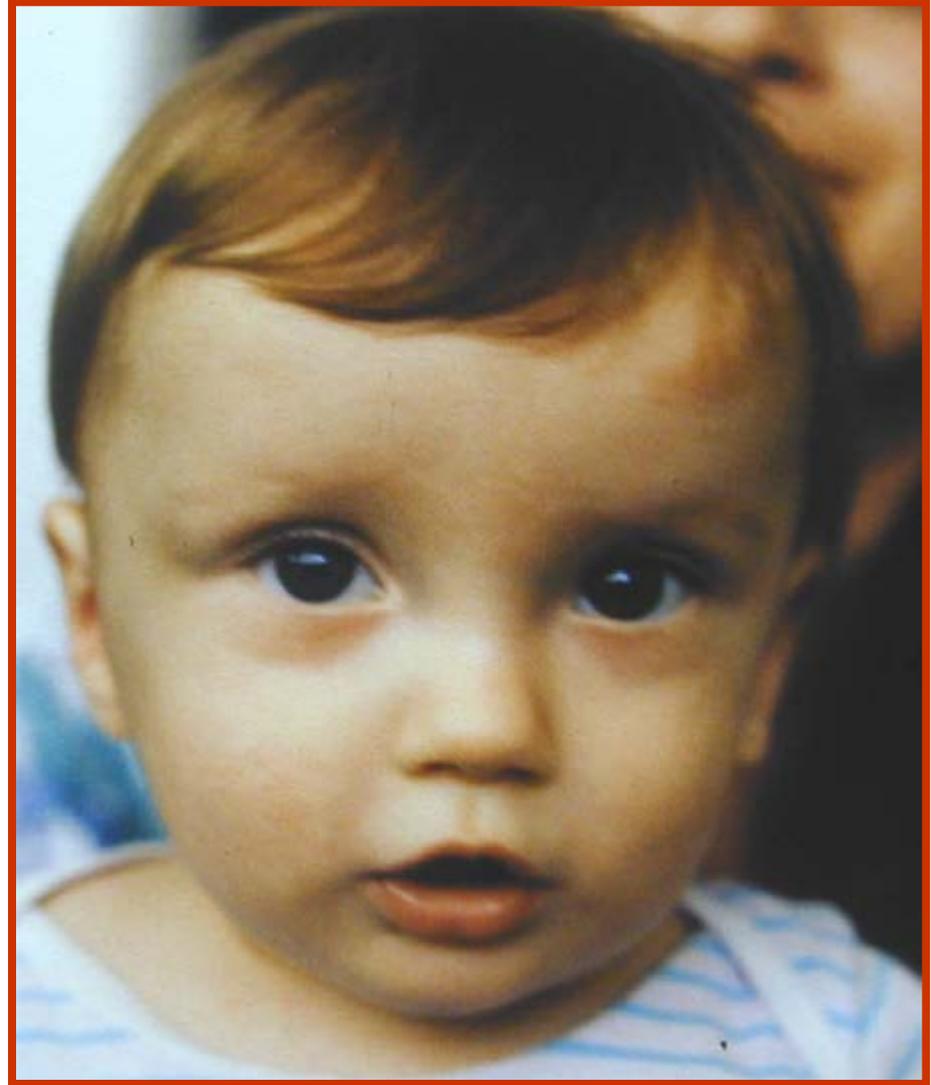
(> 6 months)

“π” technique
(squeeze)





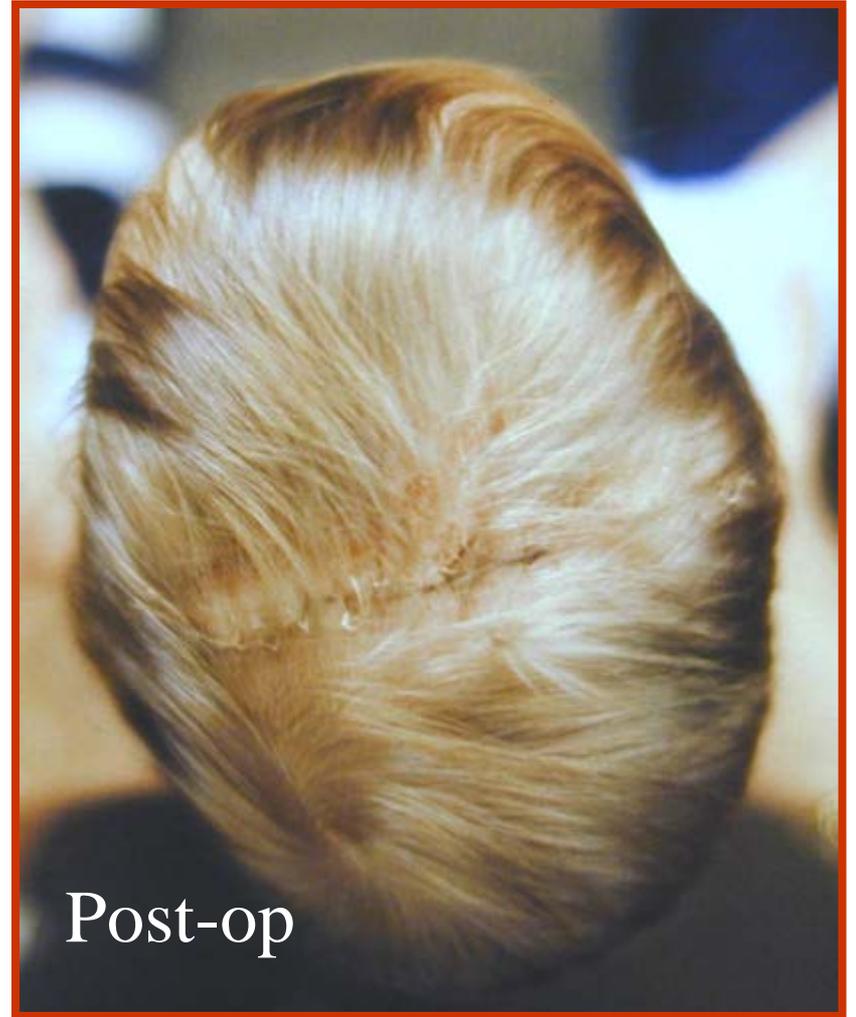
Pre-op



Post-op



Pre-op



Post-op

Scafocefalia

Approccio mini-invasivo

J Neurosurg 88:77-81, 1998

Journal of Neurosurgery: Pediatrics

Endoscopic craniectomy for early surgical correction of sagittal craniosynostosis

DAVID F. JIMENEZ, M.D., AND CONSTANCE M. BARONE, M.D.

Divisions of Neurological Surgery and Plastic and Reconstructive Surgery, University Hospitals and Clinics, University of Missouri, Columbia, Missouri

See the corresponding article in this issue, pp 407-417.

J Neurosurg (Pediatrics 5) 100:403-406, 2004

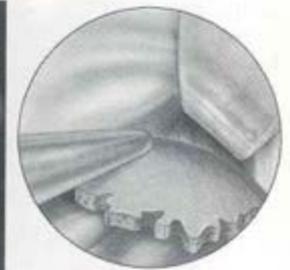
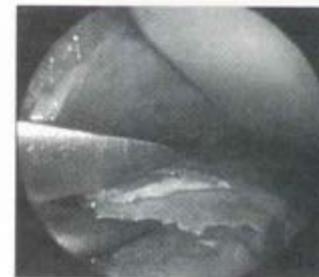
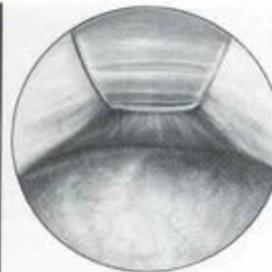
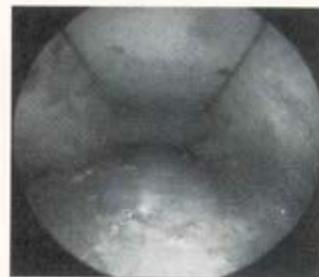


Editorial

Endoscopy-assisted craniosynostosis

JOHN PERSING, M.D.

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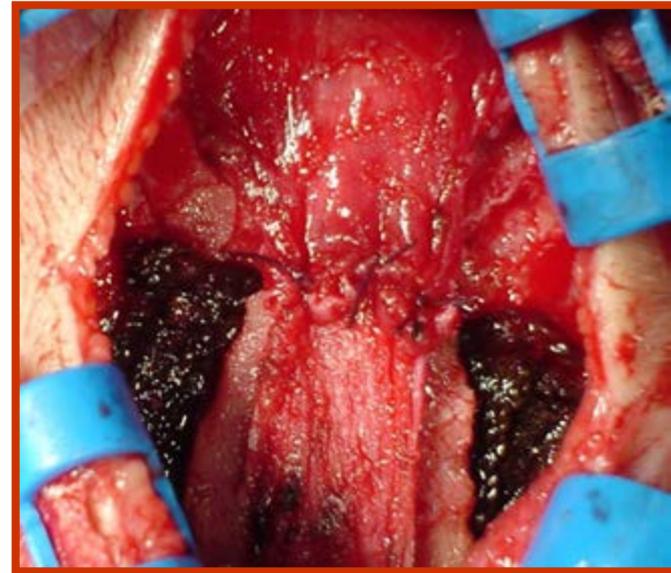
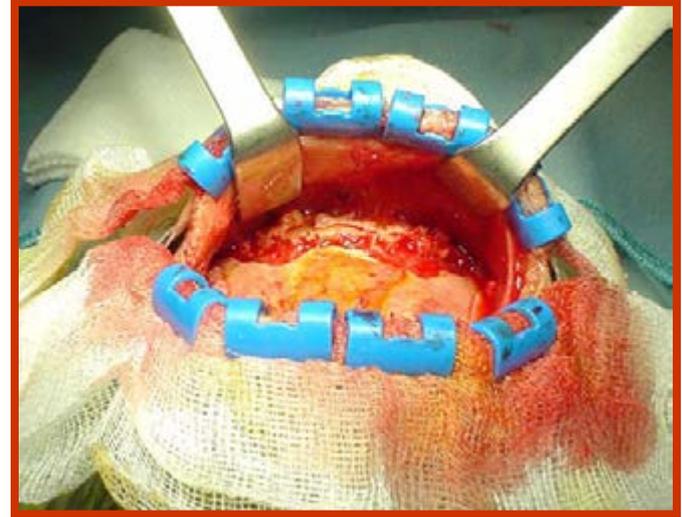


Il “nostro” approccio
mini-invasivo

(2 piccole incisioni)

32 casi (2 anni)





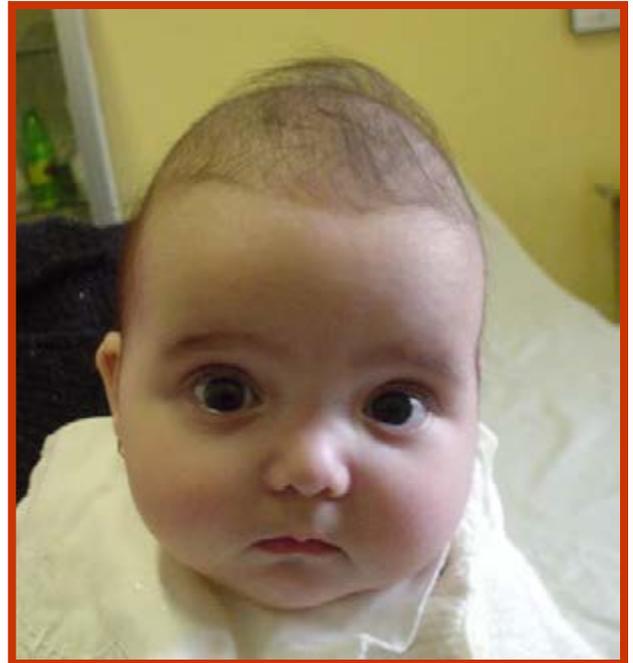


DX

DX

RX post - op



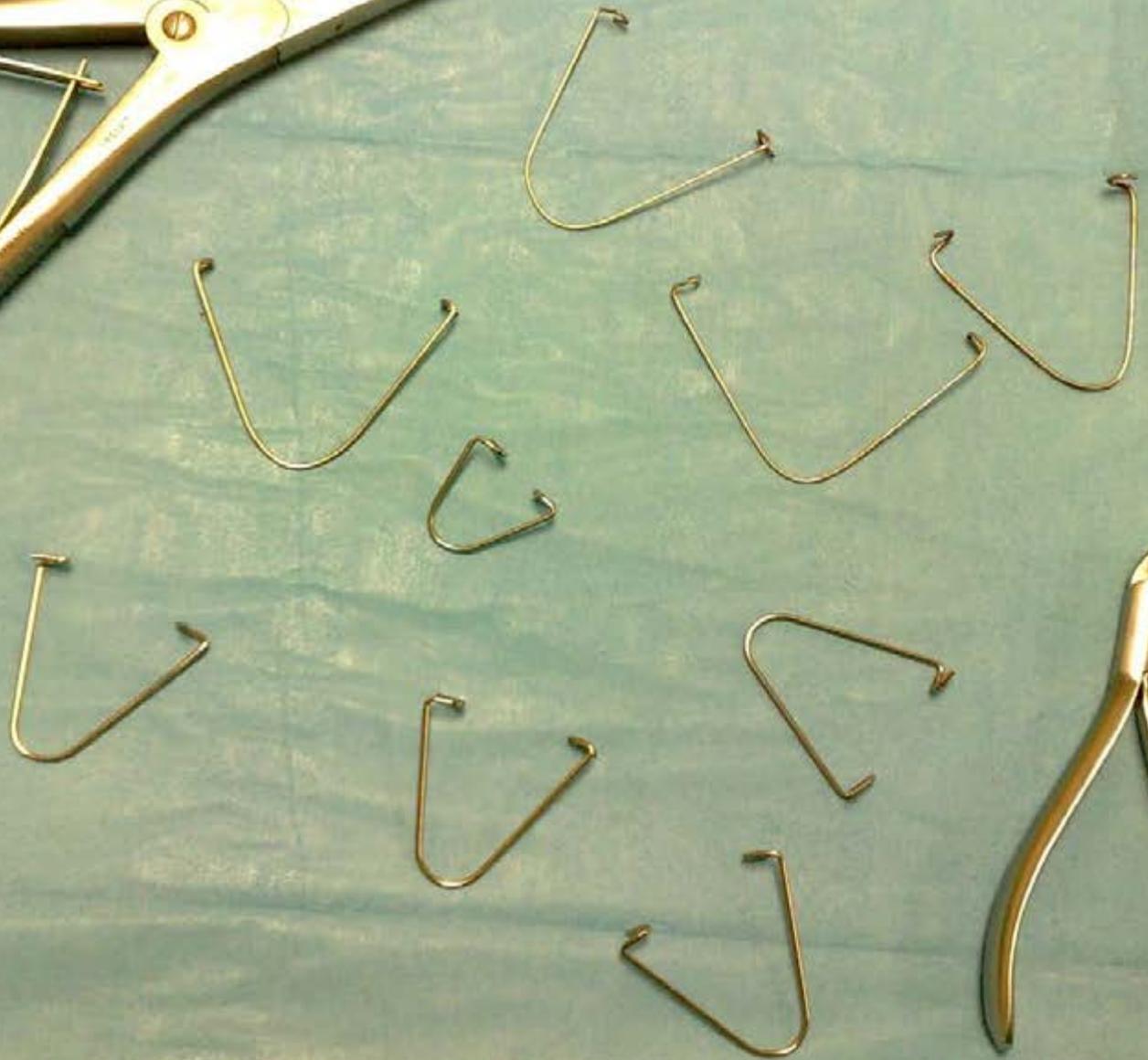


Risultato chirurgico



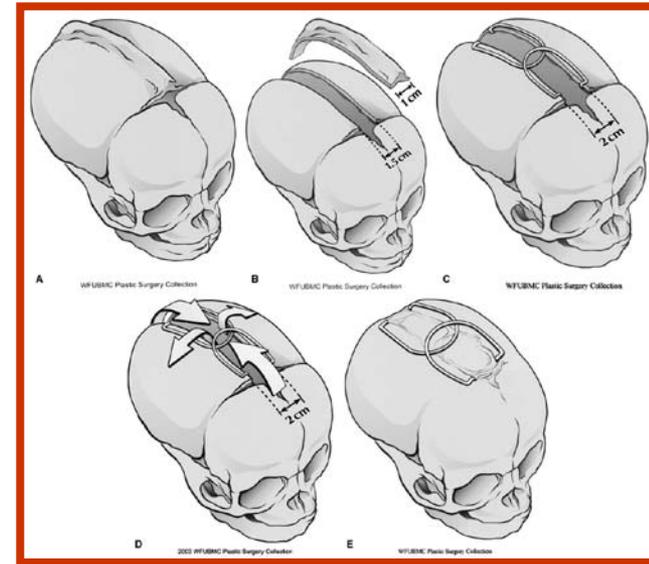
Spring Assisted Surgery (SAS)

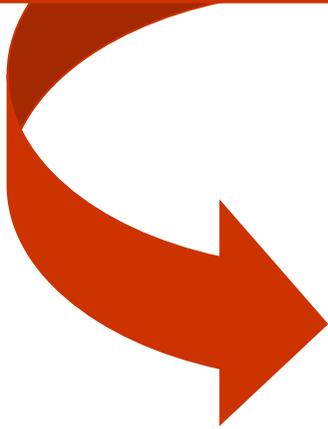
- Nuova tecnica chirurgica (Lauritzen, 1997)
- Correzione dinamica della volta cranica
- Chirurgia meno aggressiva
- Minore morbilità
- Buoni risultati estetici e funzionali
- • Chirurgia in due tempi: impianto e rimozione



Spring

- Stripping sagittale di 1 cm
- Numero medio di spring: 2
- Tempo medio di rimozione spring alla riossificazione: 4 mesi





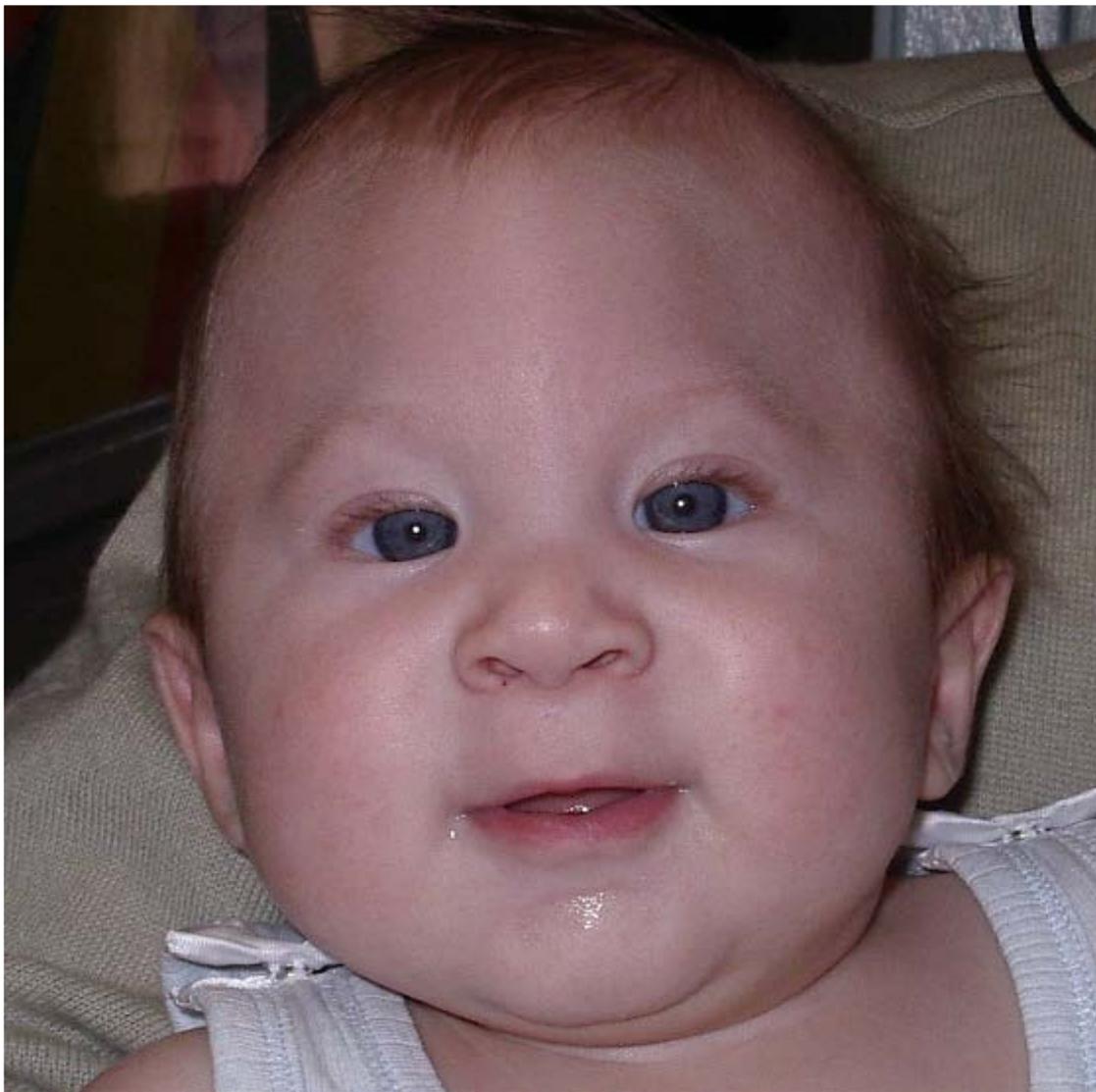
Controllo a
un anno





Controllo quattro anni dopo

Problema: necessità di un secondo intervento



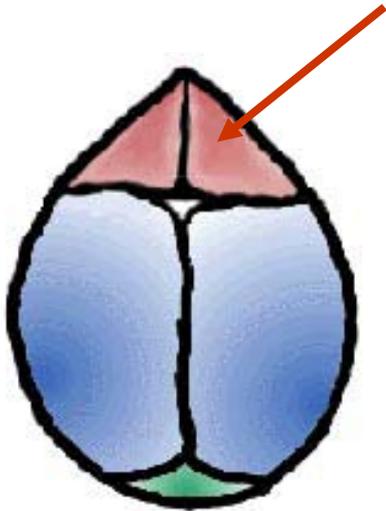
Fronte triangolare

Cresta

Trigonocefalia

- Fronte appuntita
- Ipotelorismo
- Ipoplasia sopraorbitaria
- Scarso accrescimento frontale laterale

Chiusura precoce
della
sutura metopica



TRIGONOCEFALIA

incidence at birth 1/15.000

- 14% of cranyosinostosis
- M : F ratio 3.3 : 1
- Not linked to Parents age
- Familiarity 5.6%
- AD transmission
- 38% penetration
- 7.8% twin birth (mechanical factors)
- Drugs in pregnancy (Valproic Acid)
- **associated malformations 22%**
- **chromosomal anomalies 2%**



Skull Base in Trigonocephaly

L. Genitori, S. Cavalheiro, G. Lena, C. Dollo, M. Choux

Department of Pediatric Neurosurgery, Hôpital des enfants La Timone, Marseille, France

Key Words. Craniosynostosis · Trigonocephaly · Nasion-pterional angle · Clino-pterional angle · Bipterional distance · Nasion-clinoid distance · CT, three-dimensional

Presented at the Consensus Conference
on Craniosynostoses, Rome, 4-6 May 1995

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Ocular findings in children operated on for plagiocephaly and trigonocephaly

Danièle Denis
Lorenzo Genitori
John Conrath
Gabriel Lena
Maurice Choux

Abstract Clinical examination of patients affected by plagiocephaly or trigonocephaly reveals evident malformation of the orbits, and the ocular repercussions are more pronounced when children are operated on at an advanced age. Since it is generally accepted that binocular vision is fully developed by approximately 6 months of age, a late correction of plagiocephalic or trigonocephalic skull deformities may be an obstacle to the development of normal visual function. For the present

report we investigated astigmatism and strabismus in 53 children, 39 of whom were operated on for plagiocephaly and 14 for trigonocephaly. Traction on the ocular globe induced by the bony deformation caused by the craniosynostosis may explain astigmatism and strabismus.

Key words Plagiocephaly · Trigonocephaly · Astigmatism · Strabismus

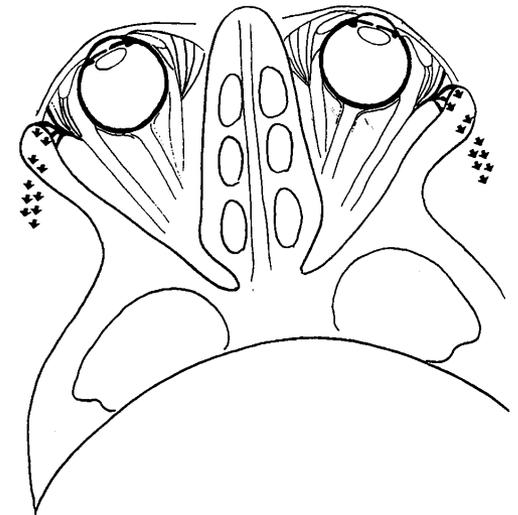
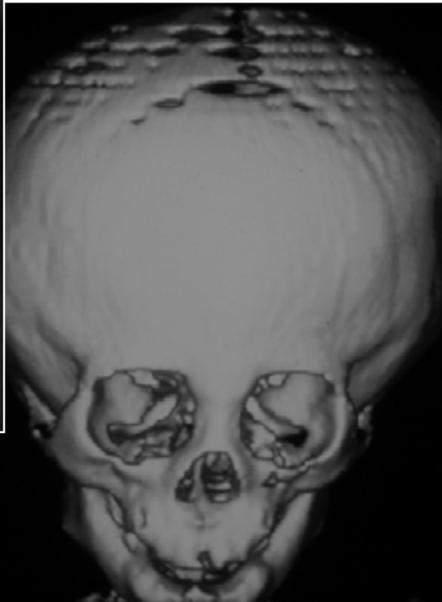
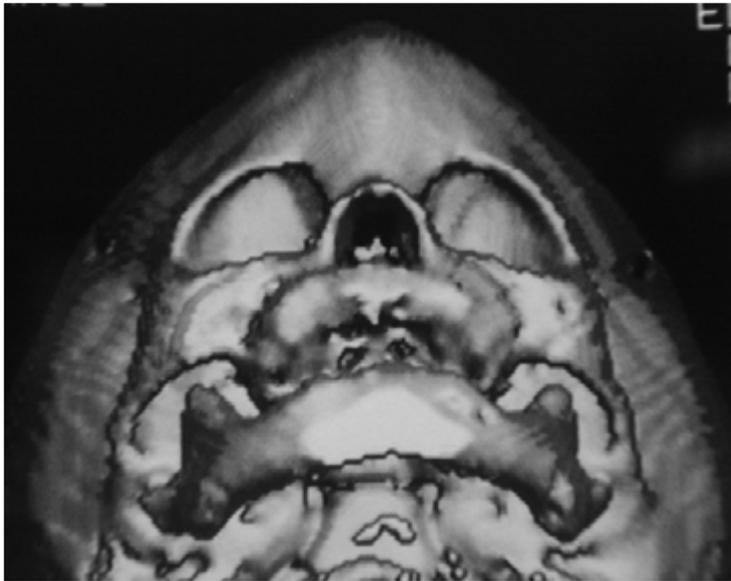


Fig. 4 Drawing of the ligament of the lateral rectus muscle inserted on the external orbital edges. Trigonocephaly bilaterally displaces the frontozygomatic region (arrows) and traction on this thick ligament is induced

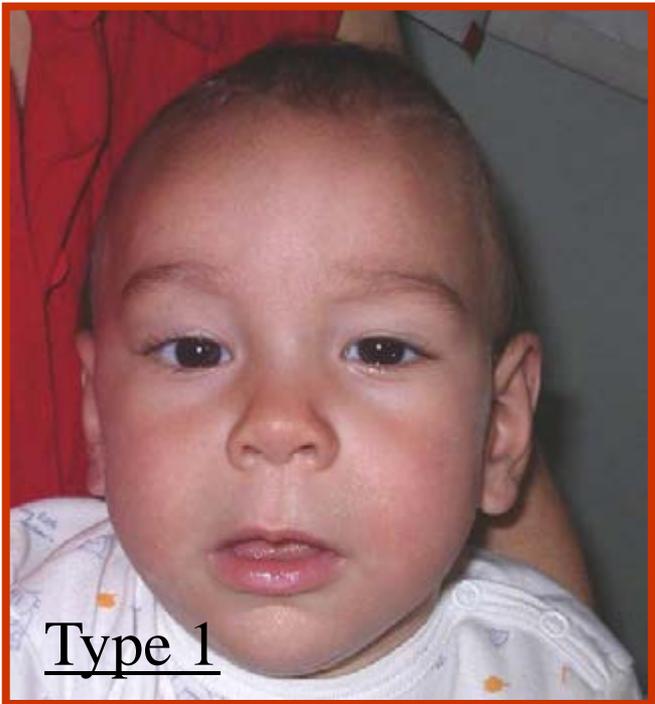
CLASSIFICATION

(Genitori, 1991)



- Type 1 early metopic synostosis;
frontal ridge
- Type 2 Type 1 +
frontal hypoplasia with temporo-parietal
expansion
orbital ridge hypoplasia
thickening and upslanting medial
walls of the orbit
back zygomatic process
- Type 3 Type 2 +
ethmoidal hypoplasia and hypothelorism

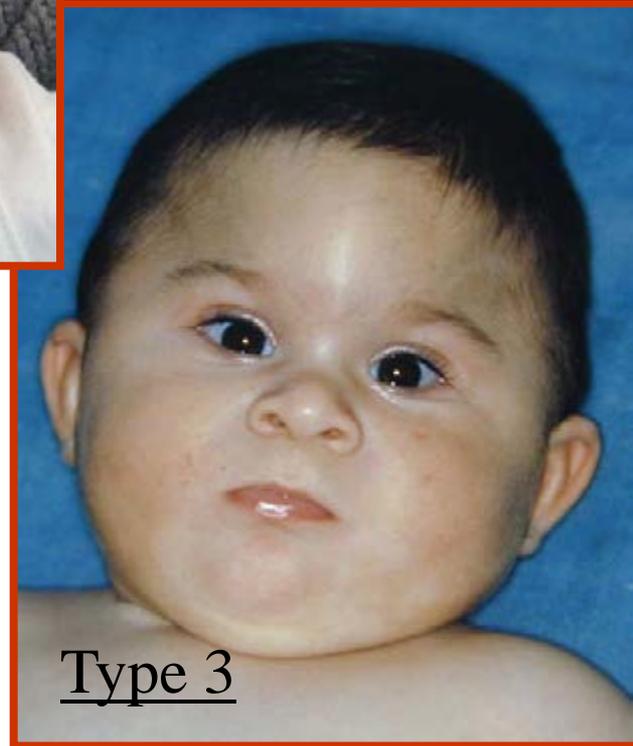
S
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G
E
R
Y



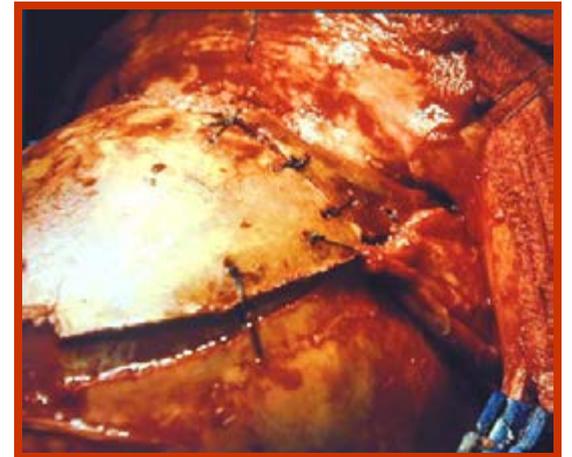
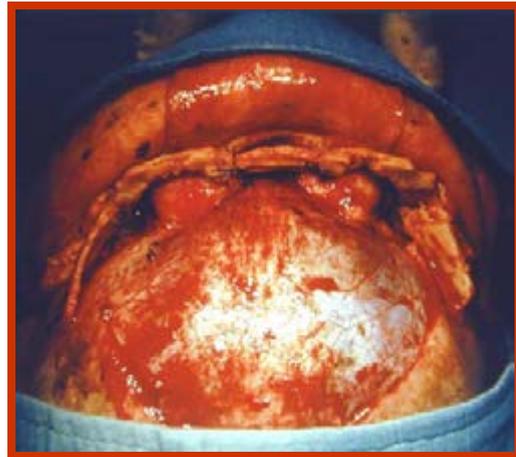
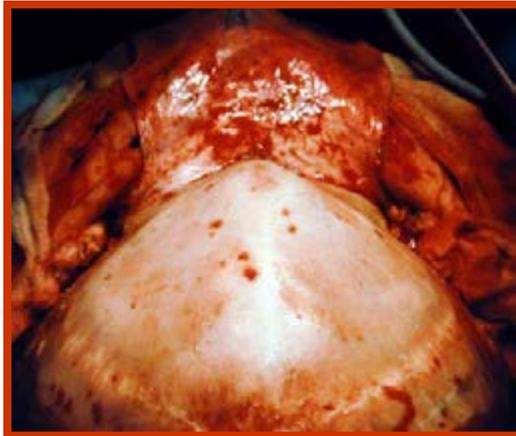
Type 1

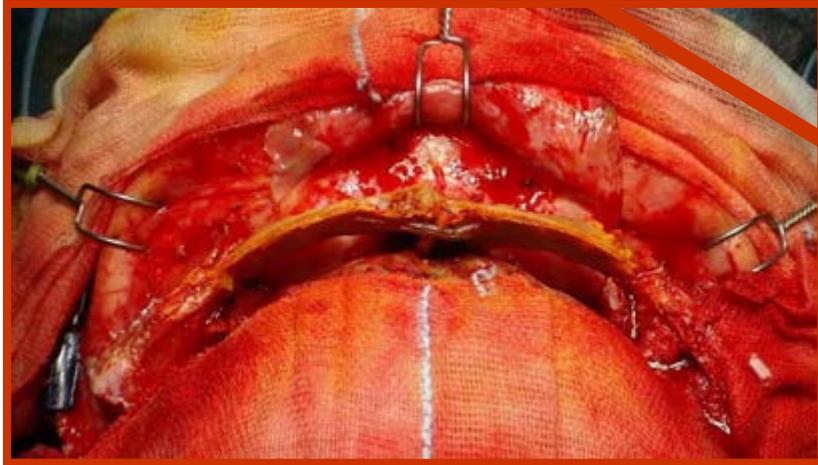


Type 2

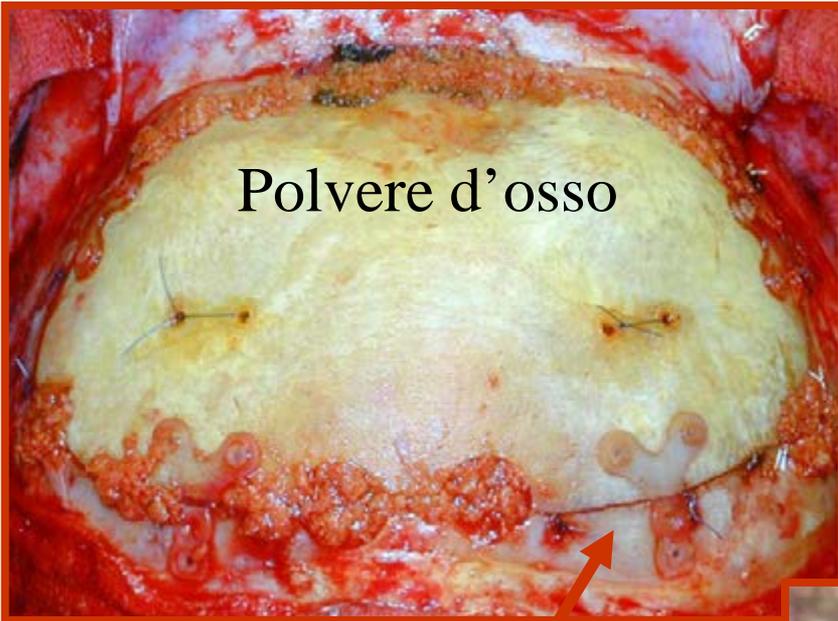


Type 3

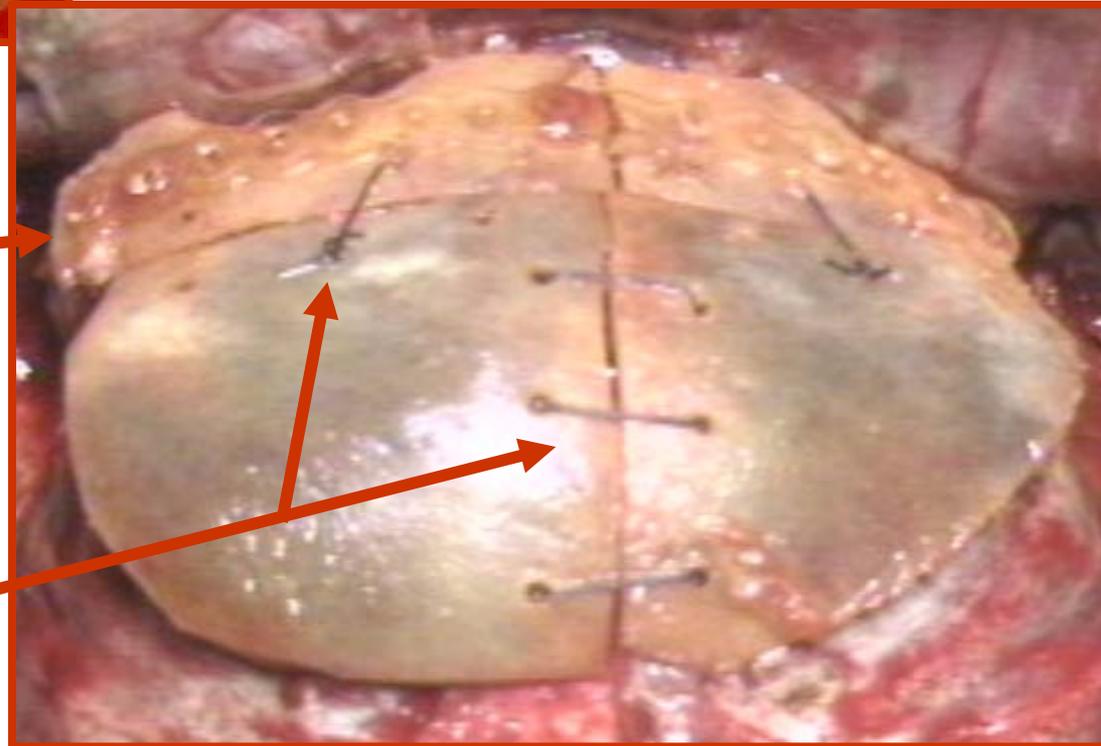




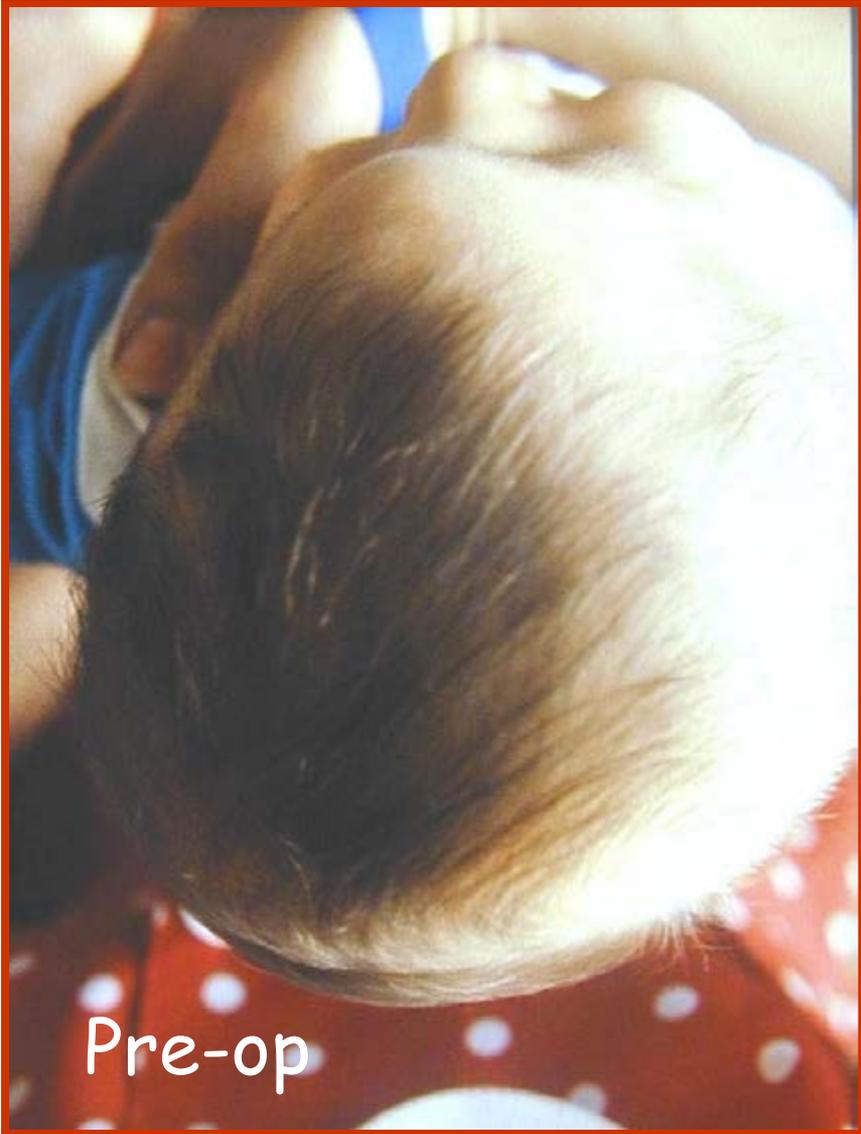
Sistemi di fissazione

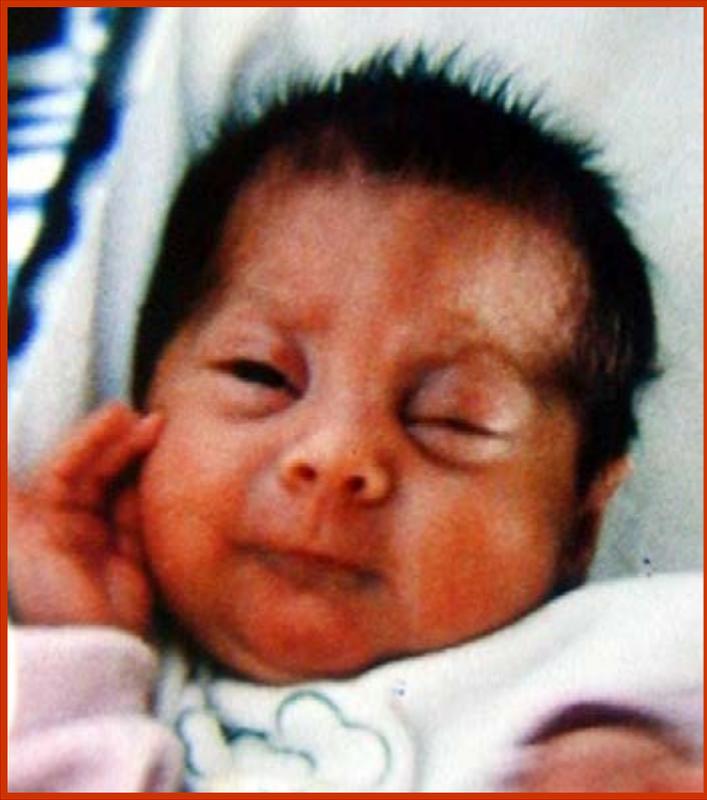


Biomaterials plates



Resorbable sutures





Pre-op

Post-op

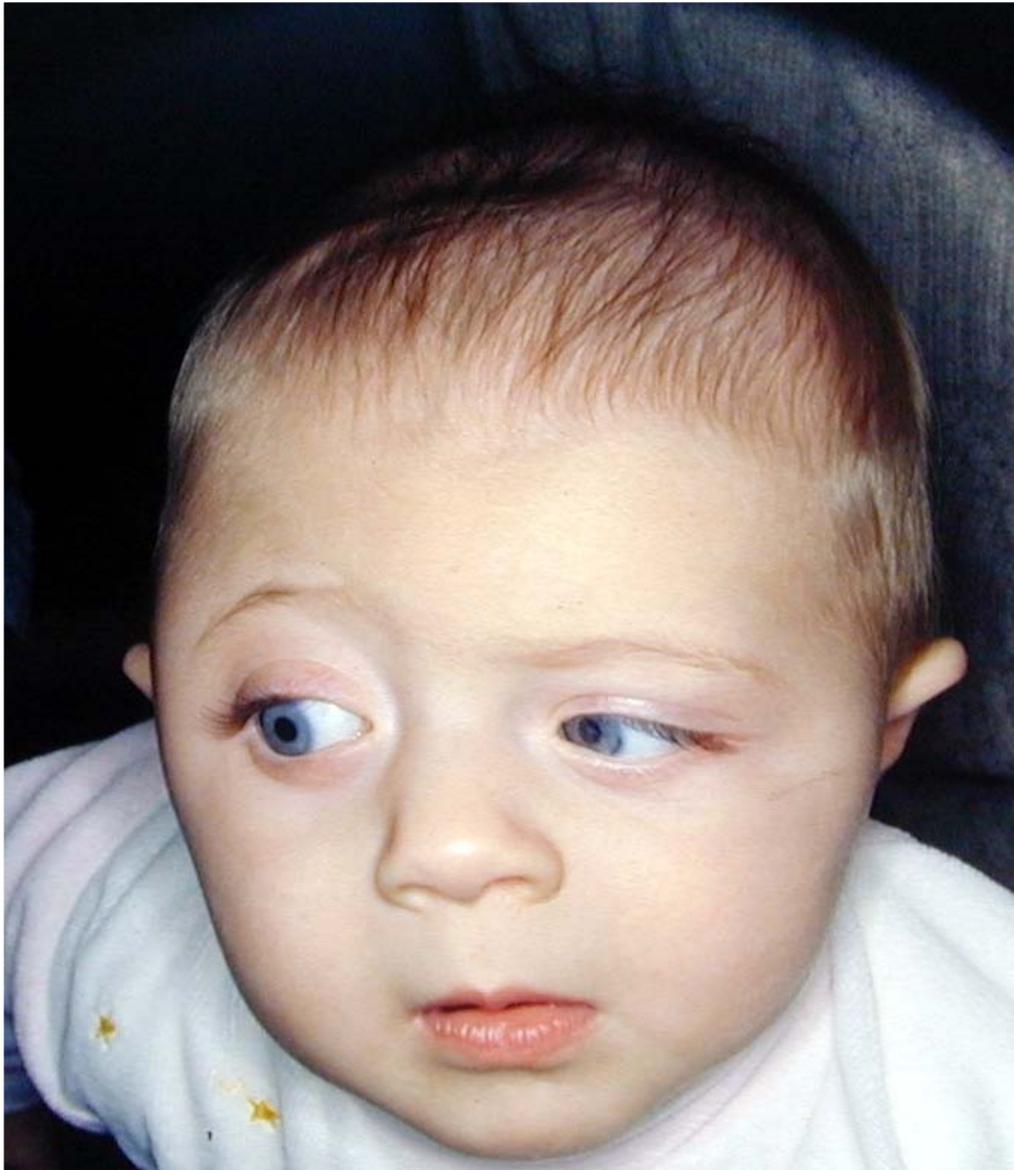




Pre-op

Post-op





Testa storta

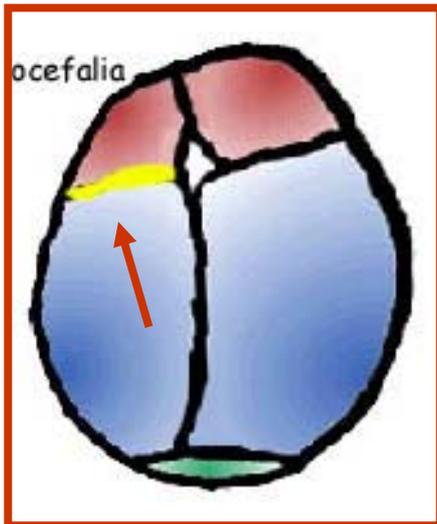
**Asimmetria
cranica**

**Asimmetria
facciale**

Plagiocefalia anteriore

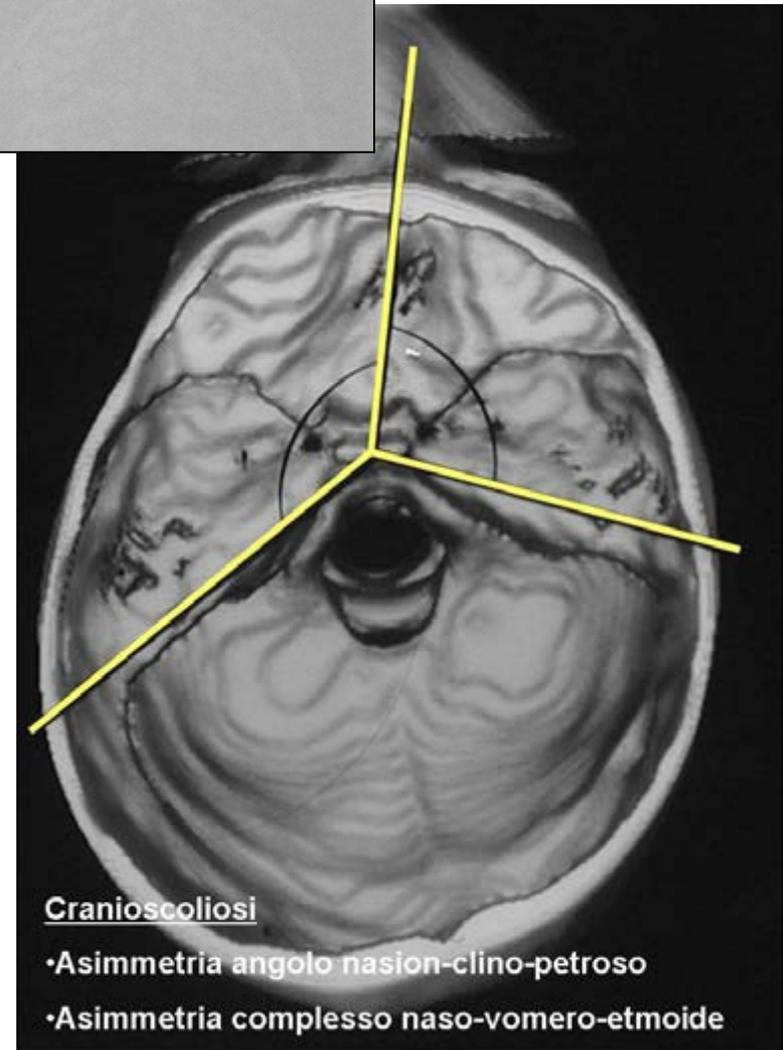
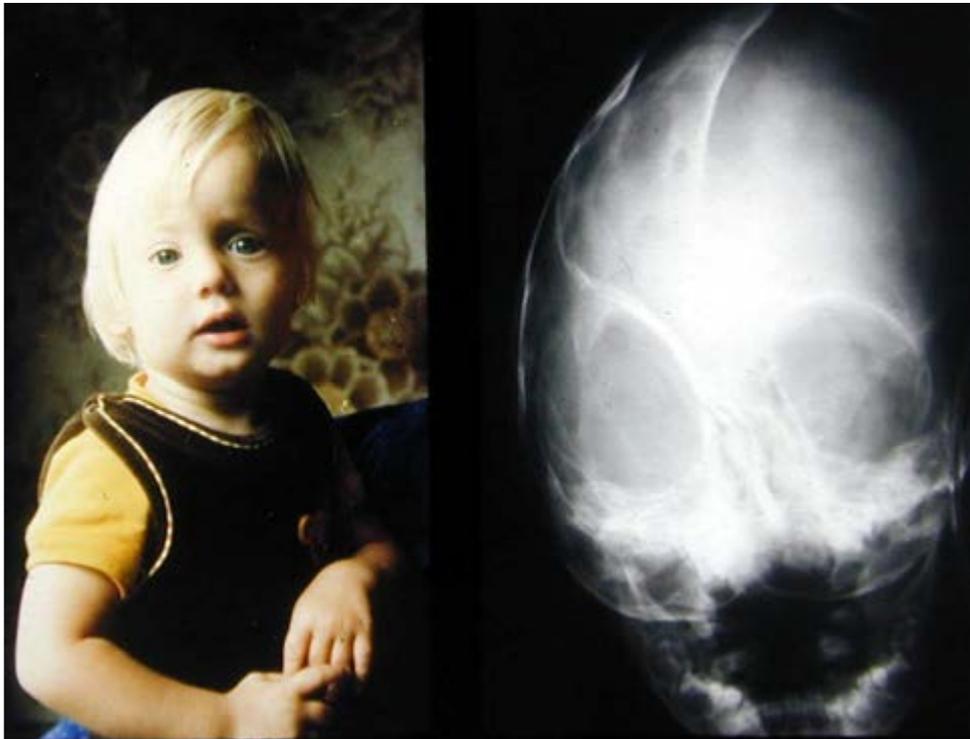
Chiusura precoce
di una
sutura coronale

- Asimmetria
- Appiattimento frontale unilaterale
- Distopia orbitaria
- Deviazione del naso
- Fronte controlaterale prominente



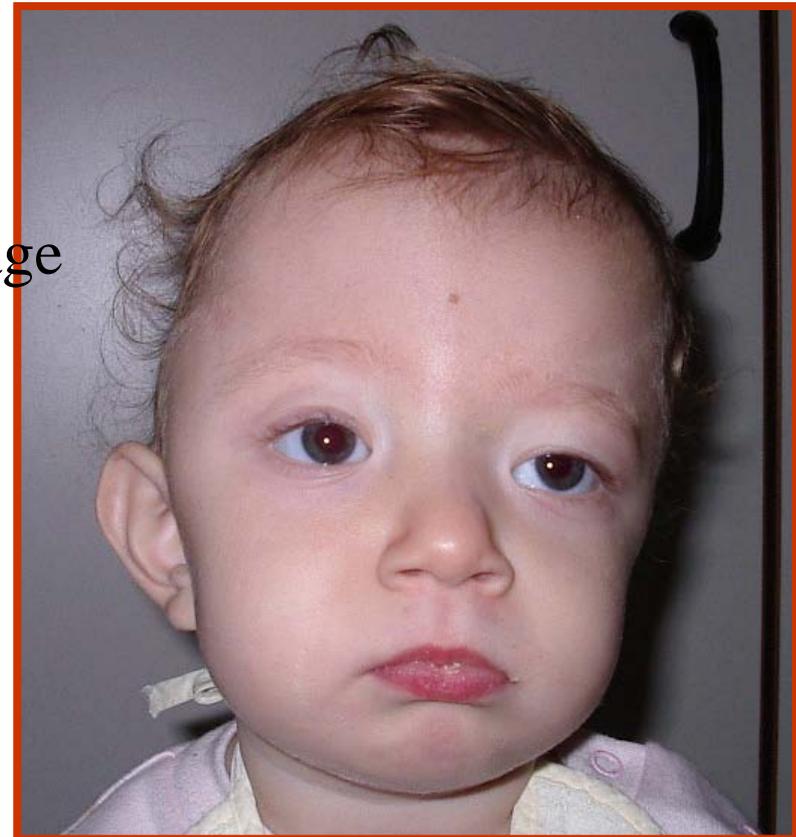
L. Genitori
N. Zanon
D. Denis
P. Erdinçler
M. Achouri
G. Lena
M. Choux

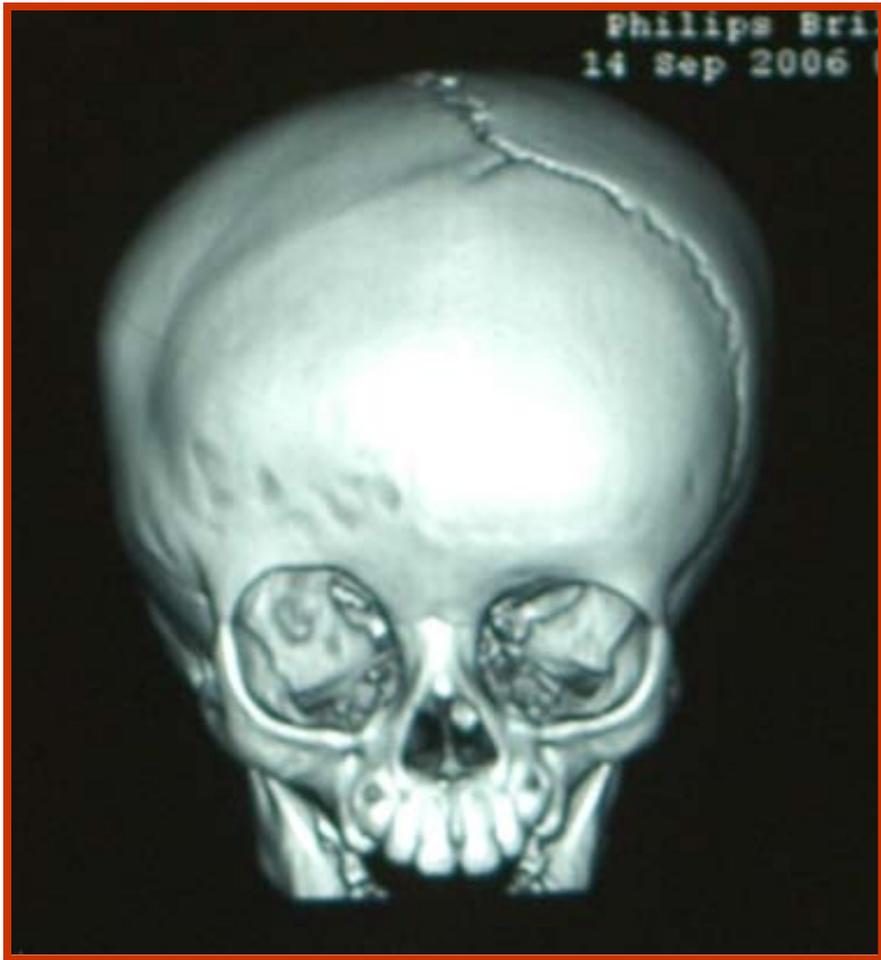
The skull base in plagiocephaly



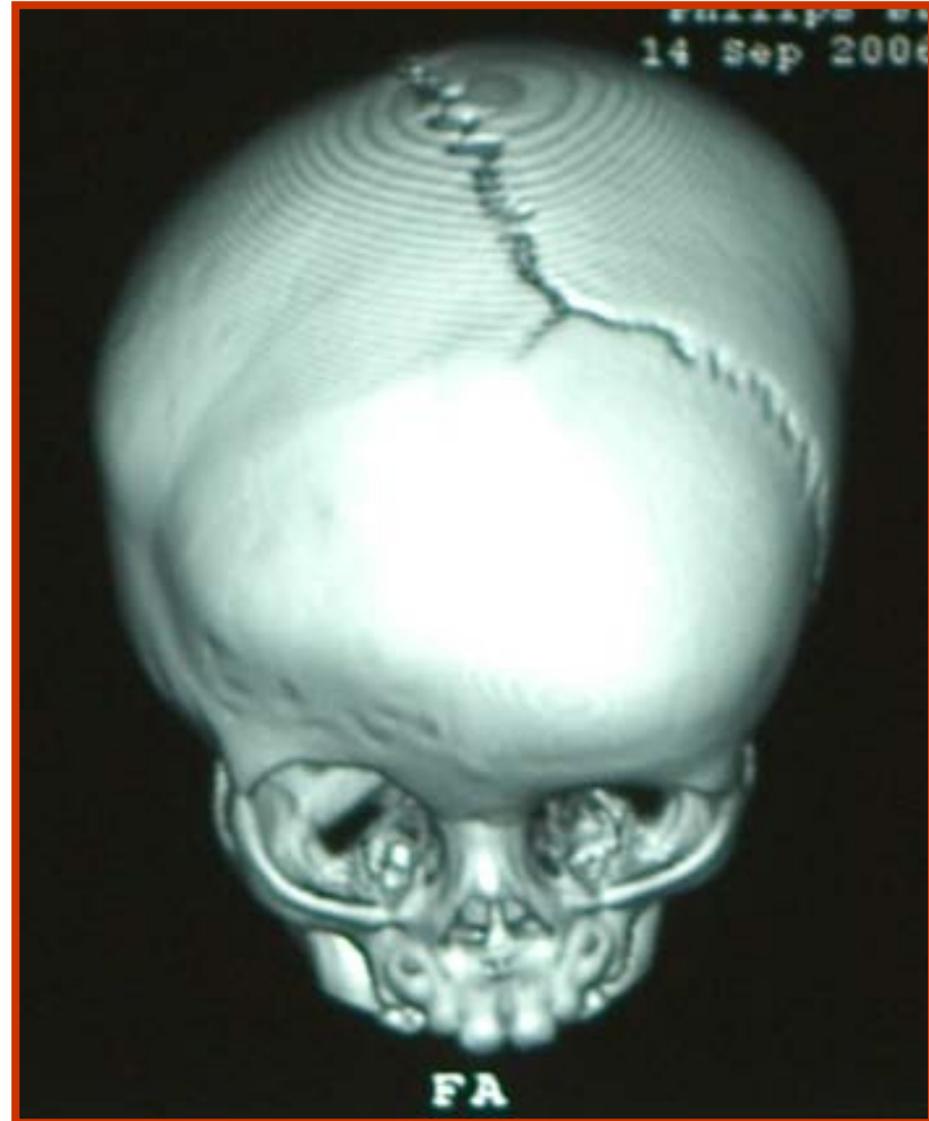
Plagiocefalia anteriore

- incidence at birth 1/16.000
- 20% of craniosynostosis
- M : F ratio 1:2
- sporadic cases linked to older parents age
- familiarity 14%
- Arms/legs anomaly possible
- FGFR3 mutations
- TWIST mutations





3D-CT scan

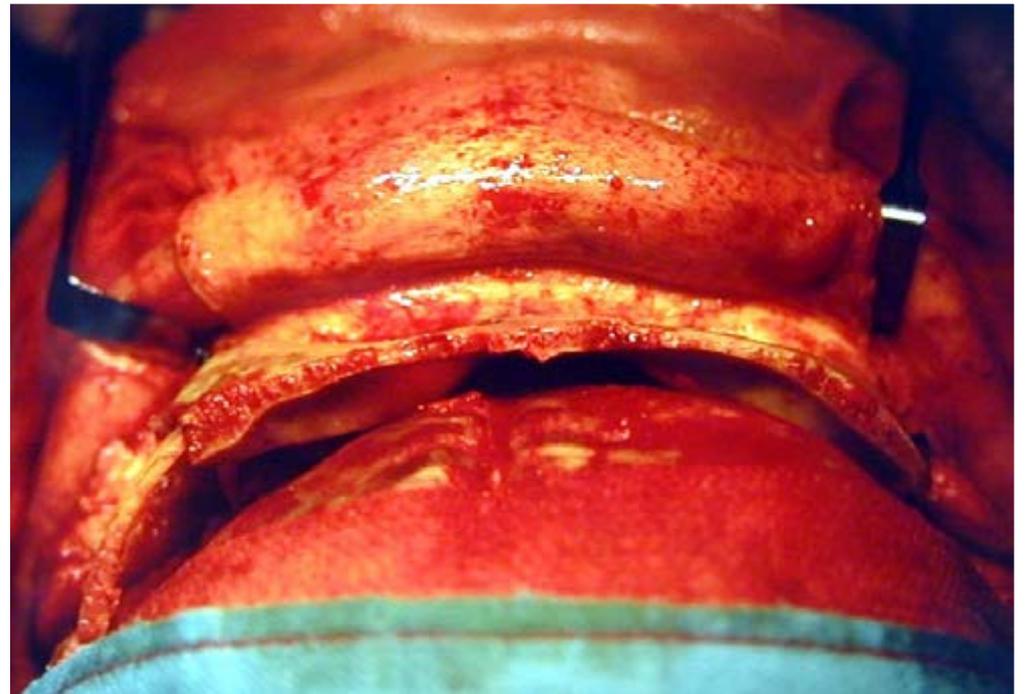
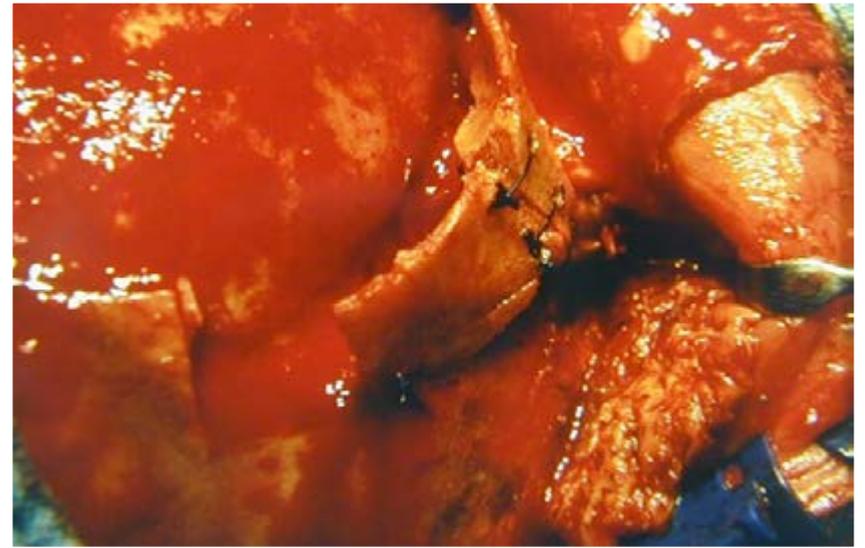


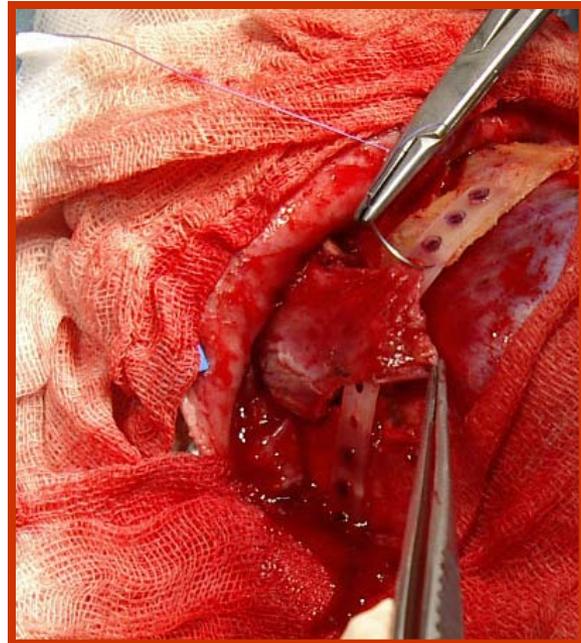
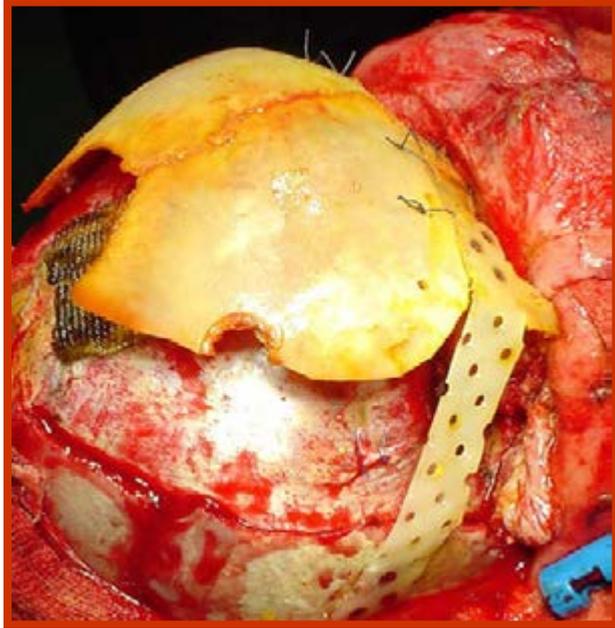
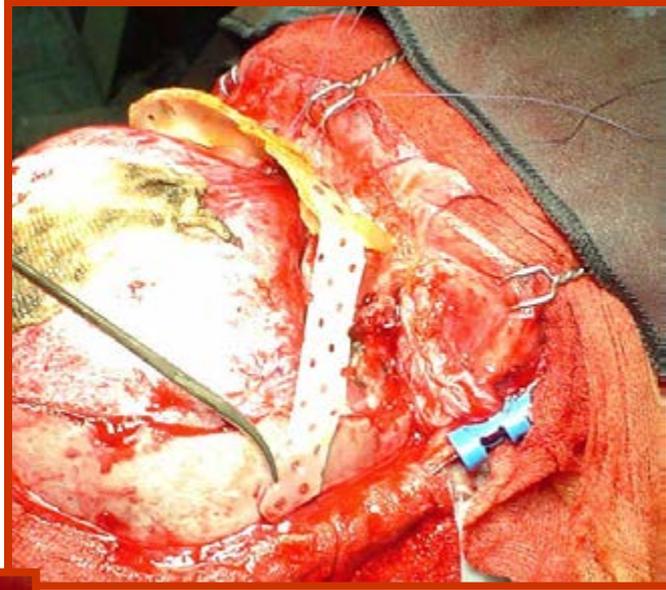
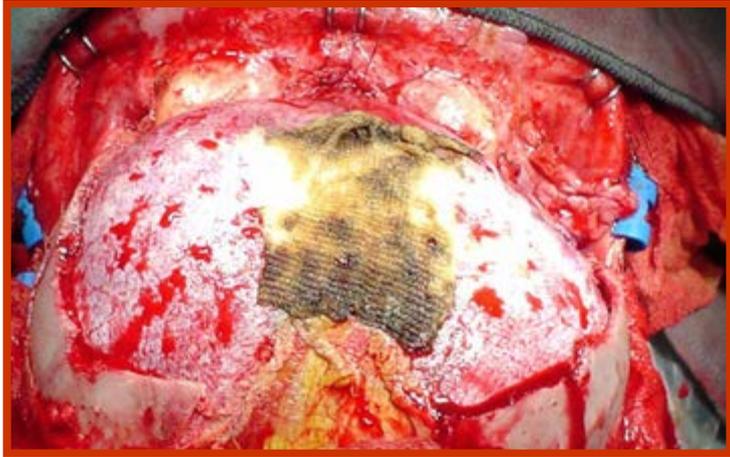
“Tilting” della testa

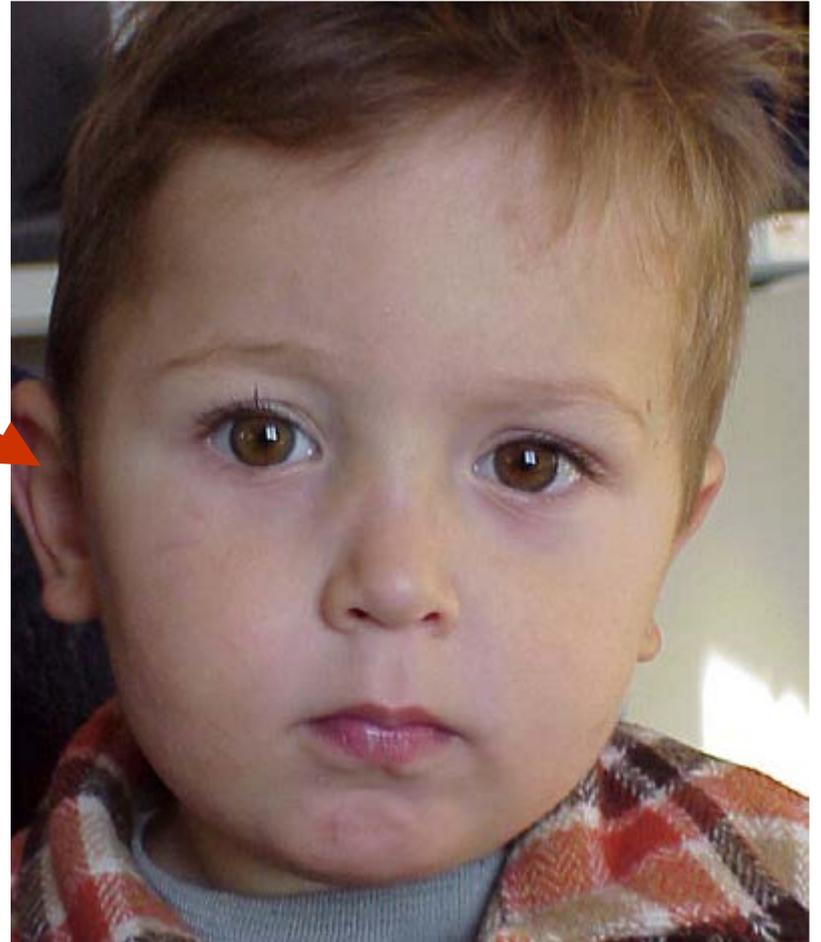
Asimmetria dei muscoli oculari

Asimmetria della giunzione occipito cervicale







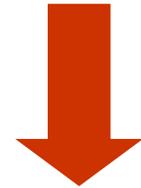
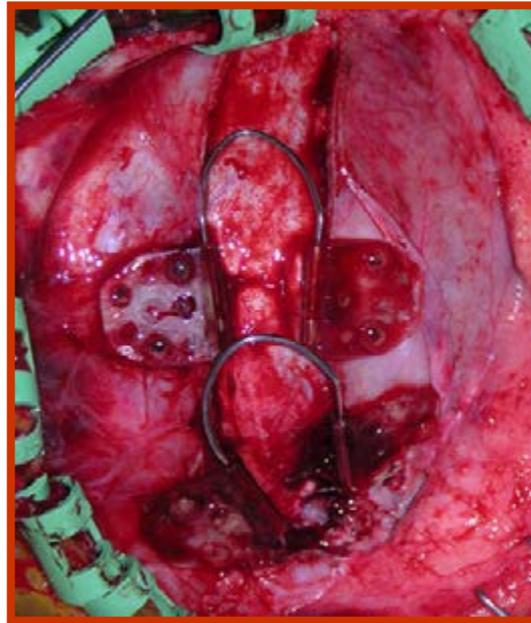
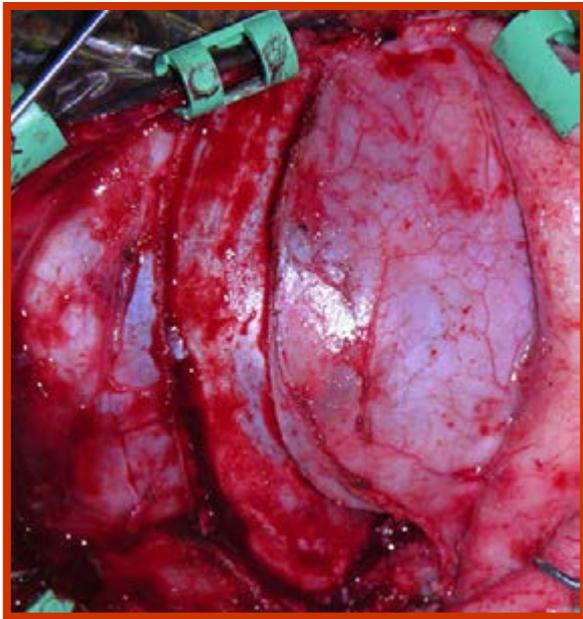


follow-up a 2 anni



follow-up a 4 anni

Spring assisted surgery Plagiocefalia anteriore





Testa piatta

Naso a sella

Turricefalia

BRACHICEFALIA



Ipoplasia bifrontale

Fronte alta

Teleorbitismo

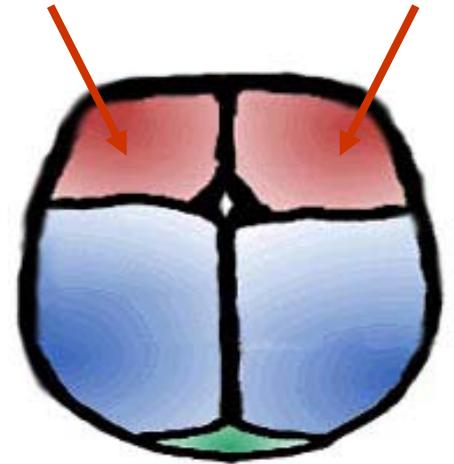
Diminuzione angolo naso frontale

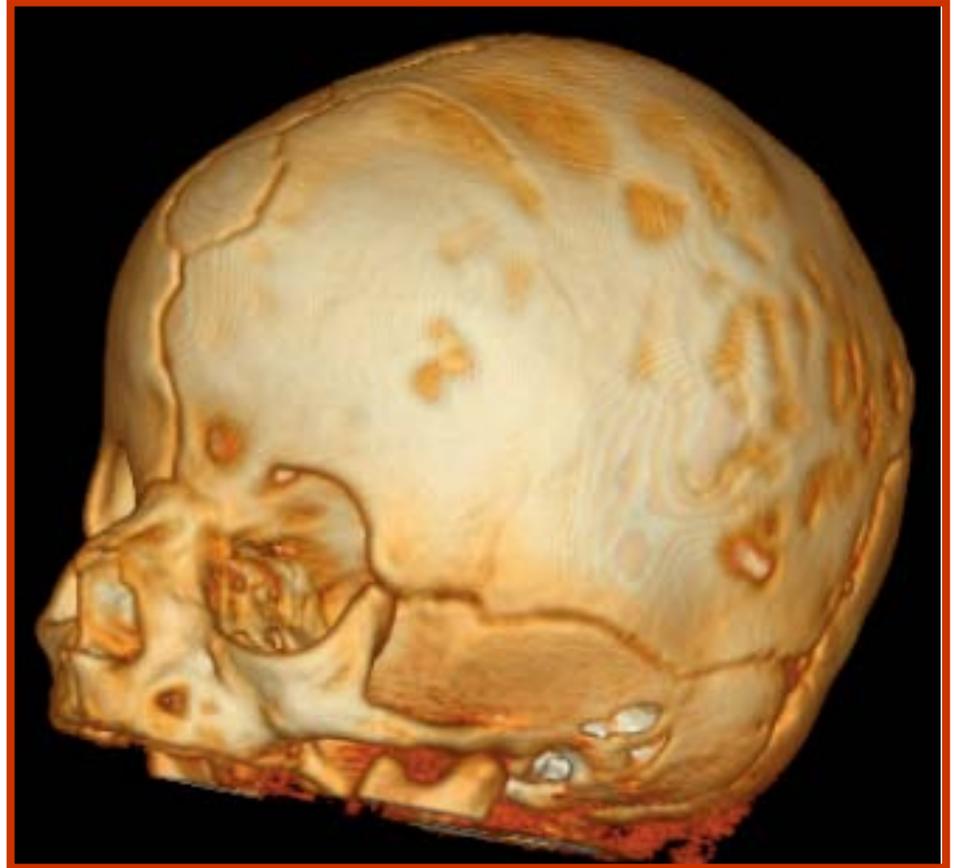
Cranium bifidum

Riduzione volume intracranico



Chiusura precoce
delle **suture coronali**



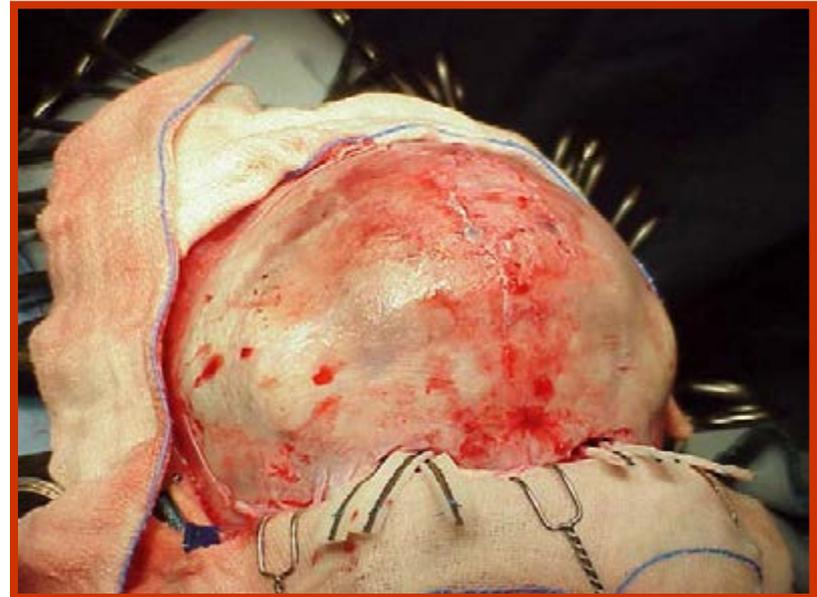


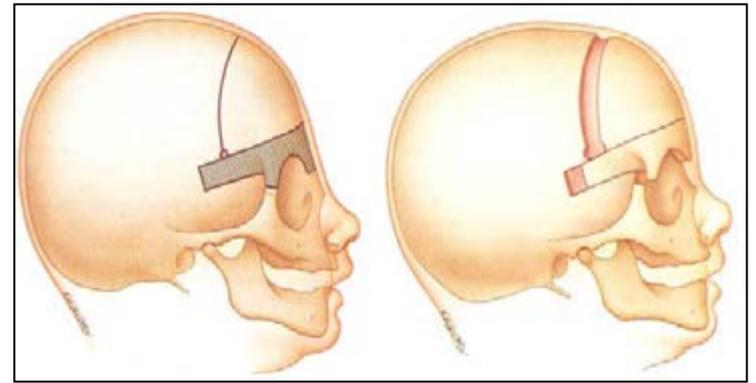
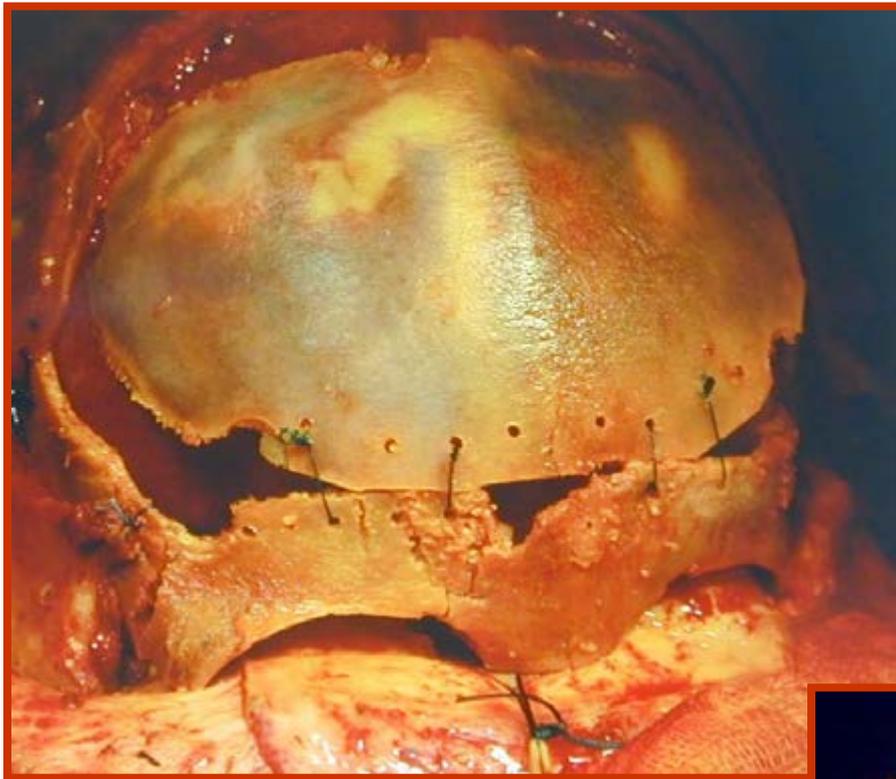
Principes du traitement

Age idéal : 4-6 mois

- Deconnection fronto orbitaire
(Front Flottant)
- Avancement fronto orbitaire
(Système auto contentif)

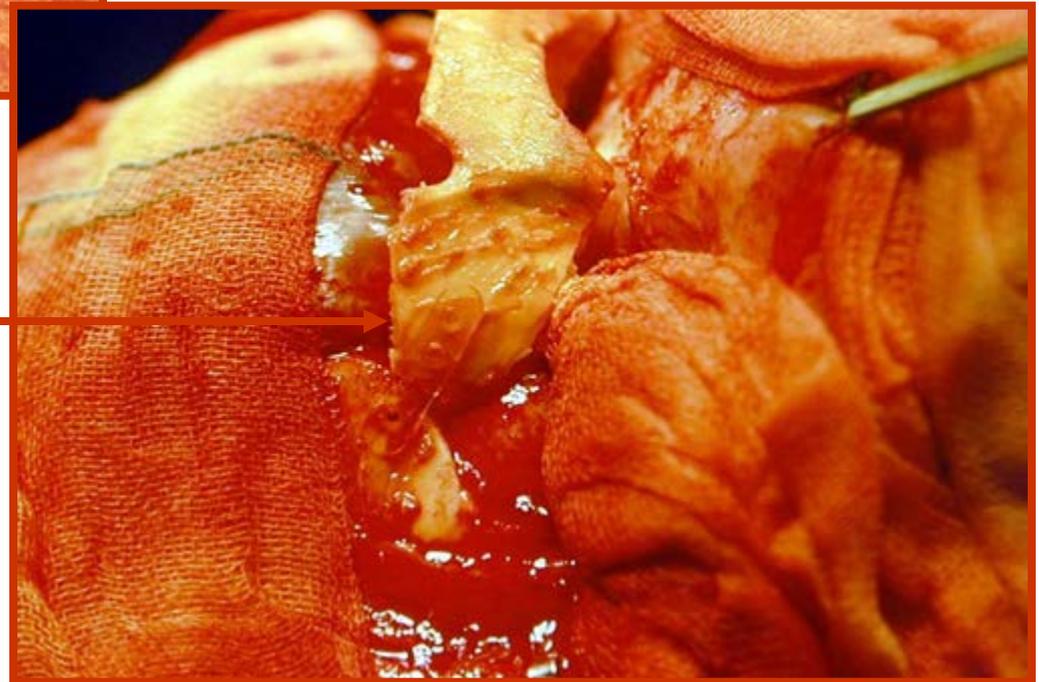
Correction de l'hypertélorisme (rare)





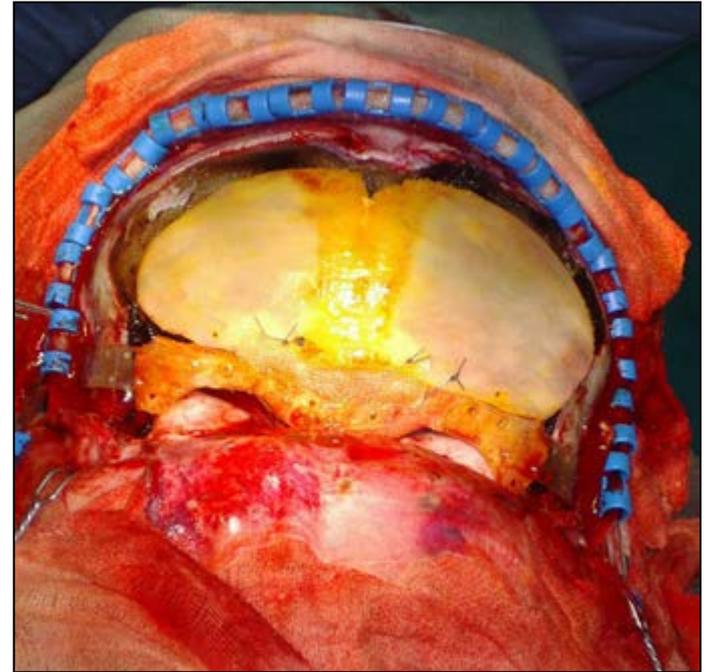
Avanzamento fronto-orbitario

Placche e viti
riassorbibili



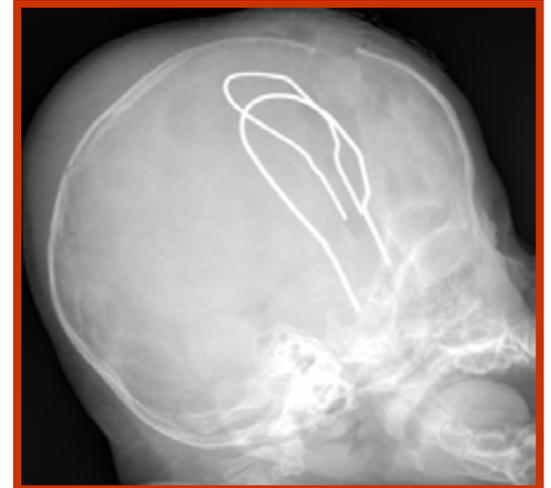


DP, 6 mesi
Brachicefalia



3 mesi post-op dopo
avanzamento fronto-
orbitario con placche

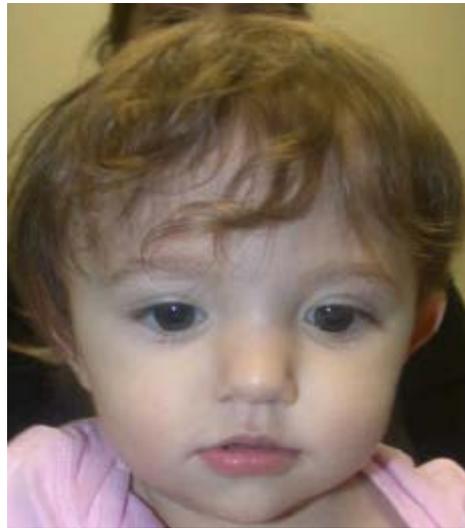
Spring assisted surgery Craniostenosi bicoronale



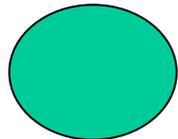
Spring assisted surgery Craniostenosi bicoronale



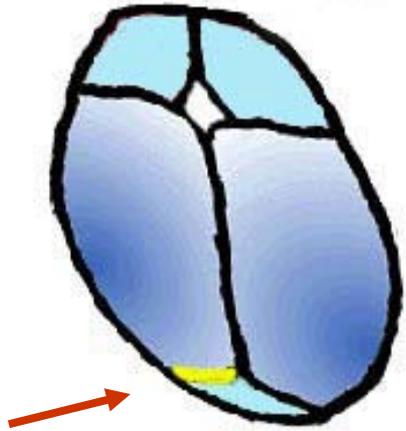
5 mesi post SAS



11 mesi post SAS



Problema: necessità di un secondo intervento

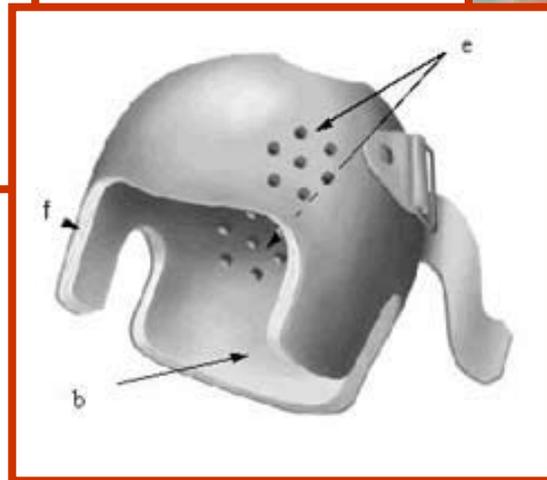
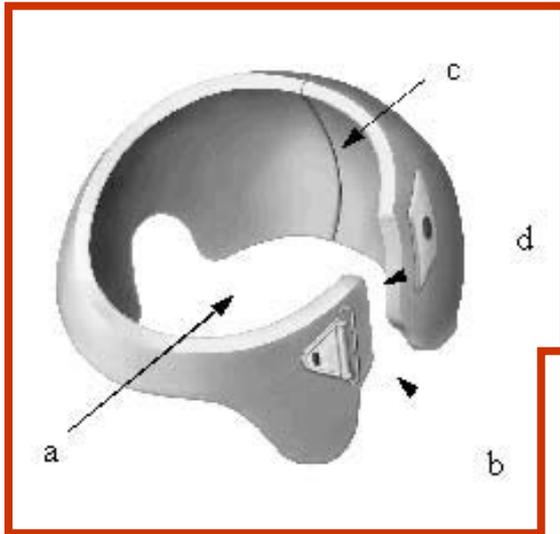
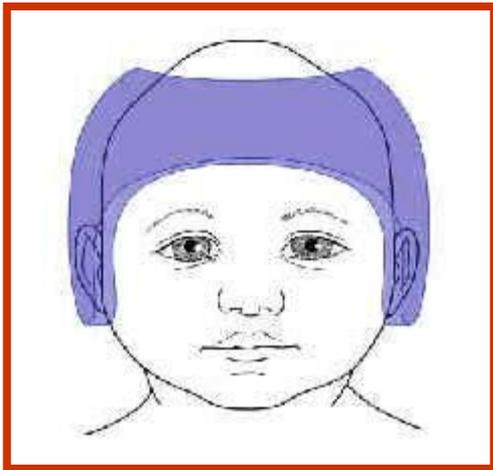


PLAGIOCEFALIA POSTERIORE

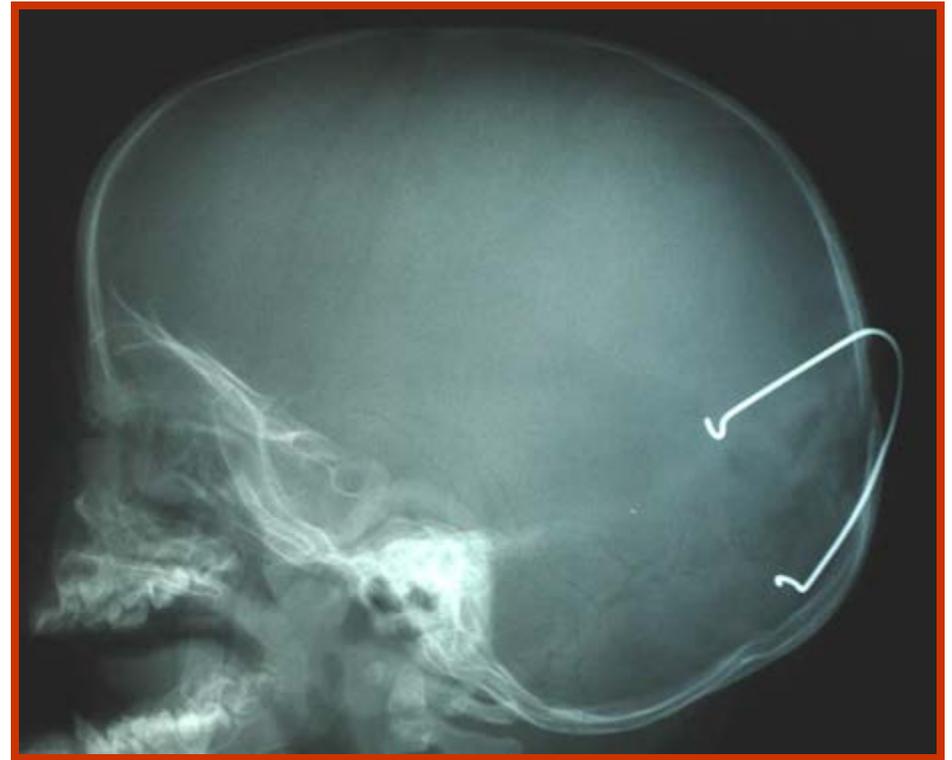
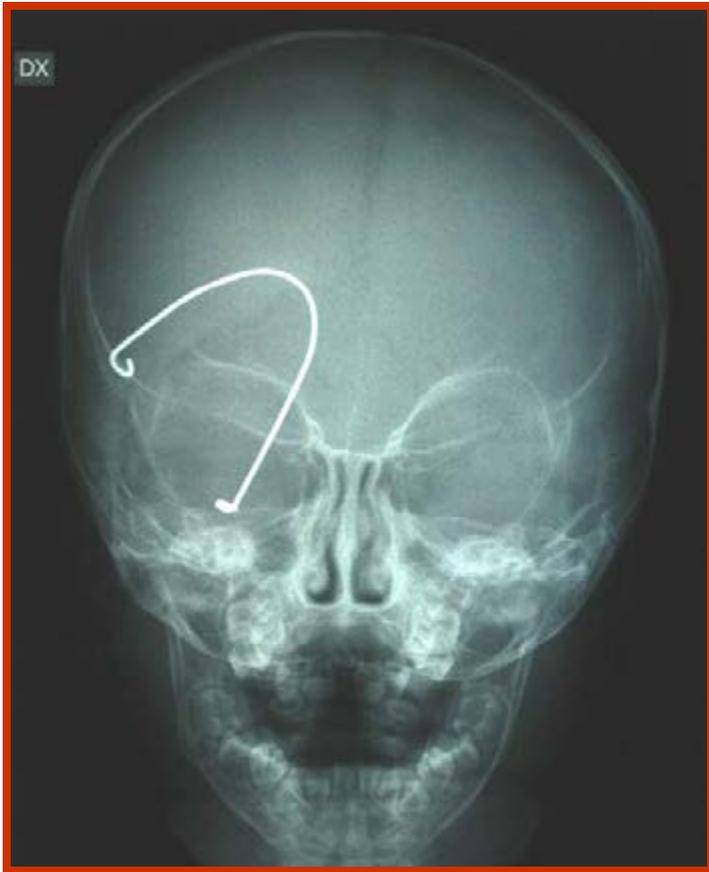
- Posizionale
- Vera craniosinostosi
- Non deficit funzionali
- Impatto estetico modesto



Plagiocefalia posteriore posizionale

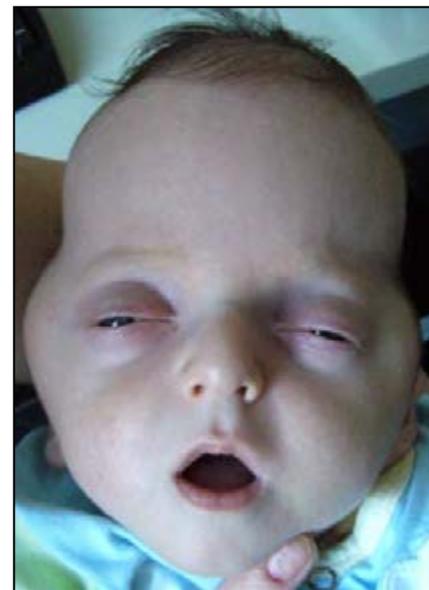


Plagiocefalia posteriore stenotica



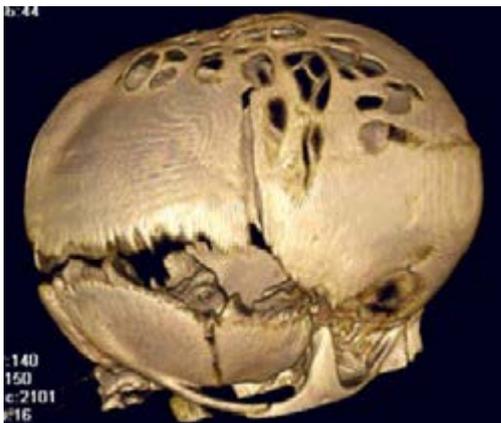
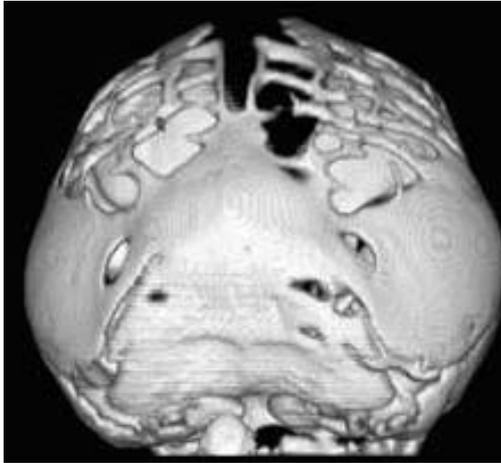


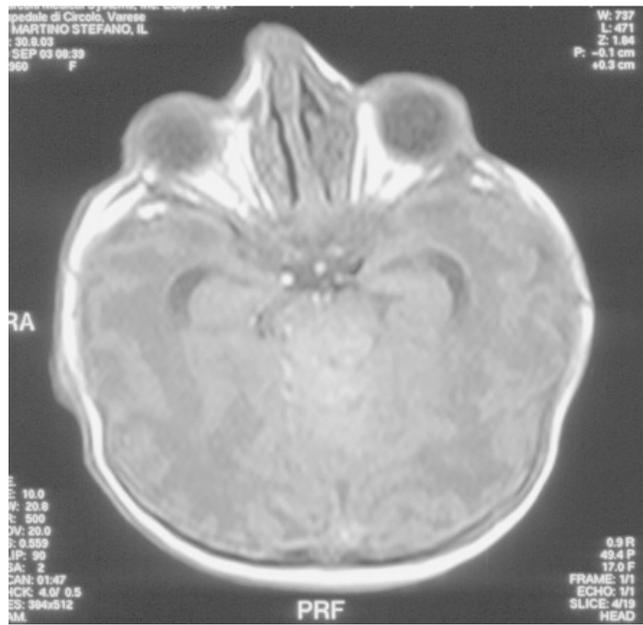
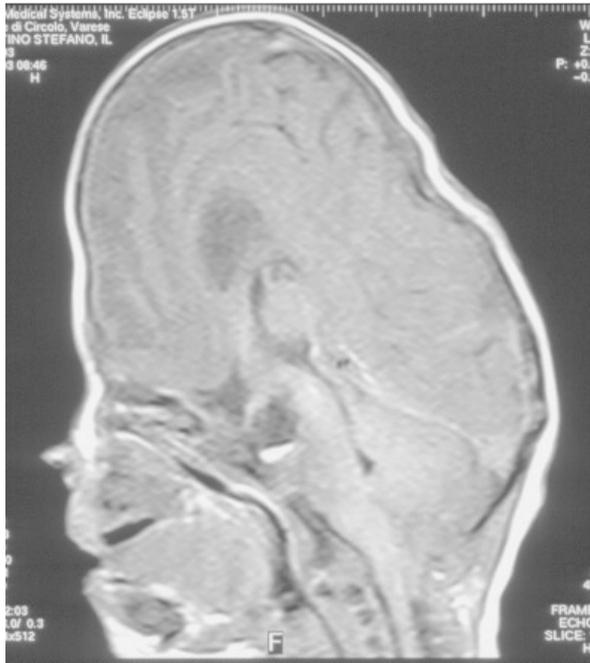
Forme “decisamente” più complesse

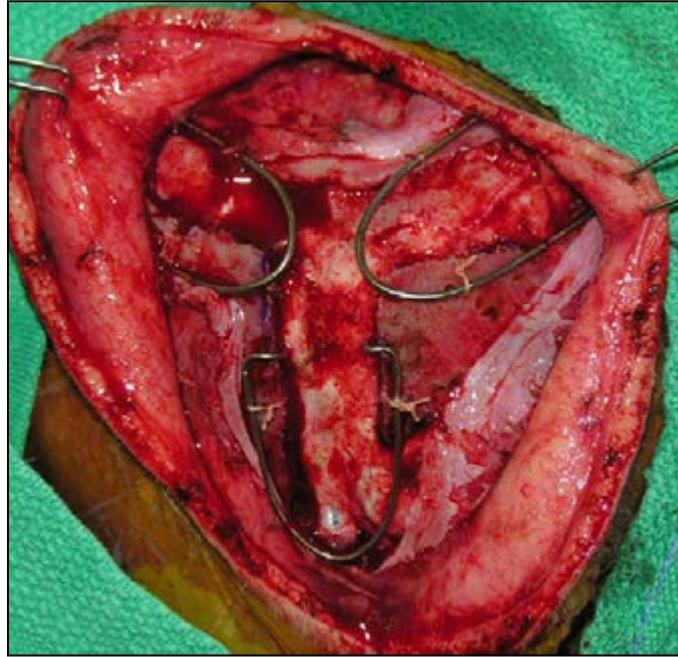
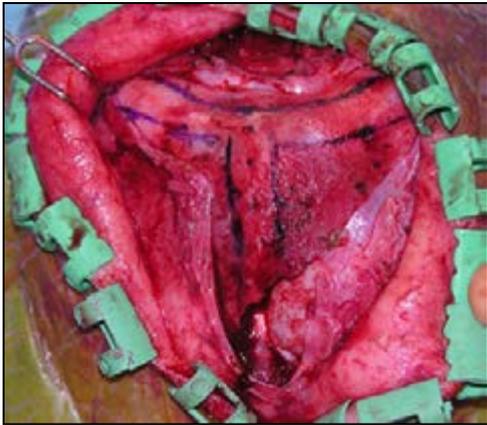
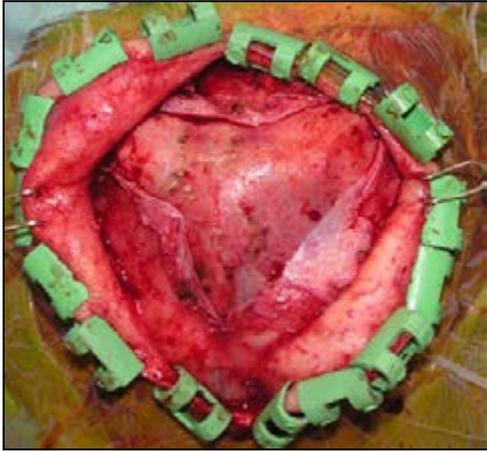


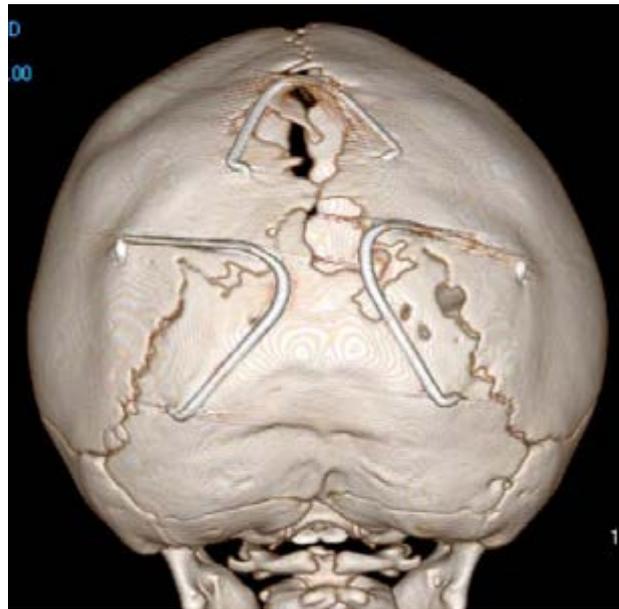
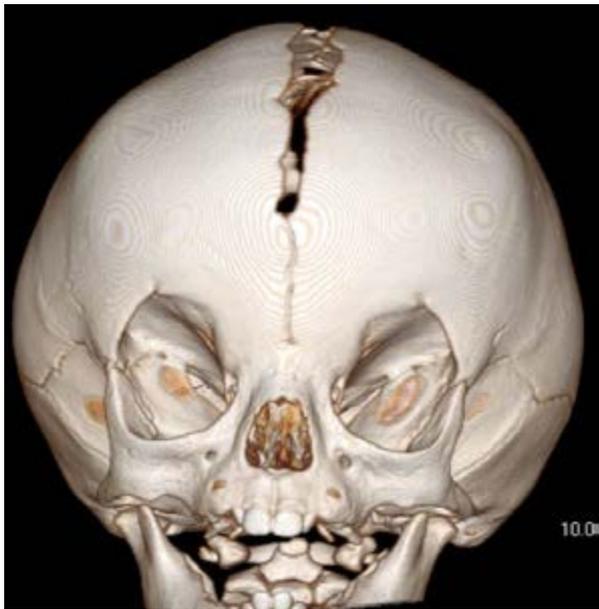
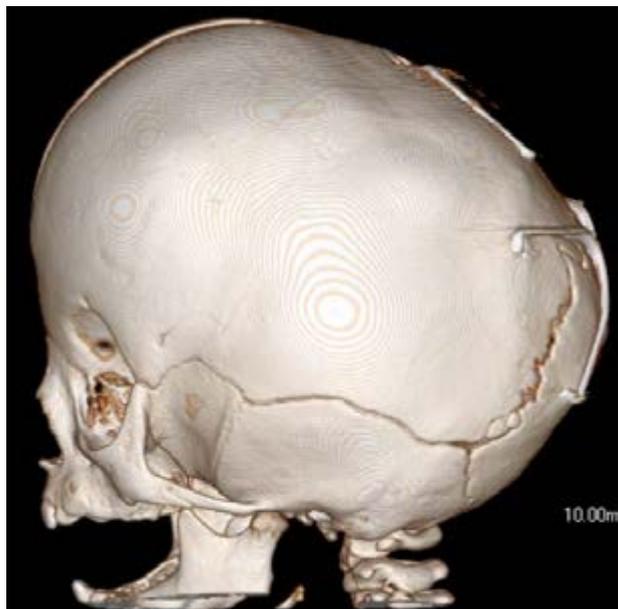
Kleeblattschadel

“Cranio a trifoglio”









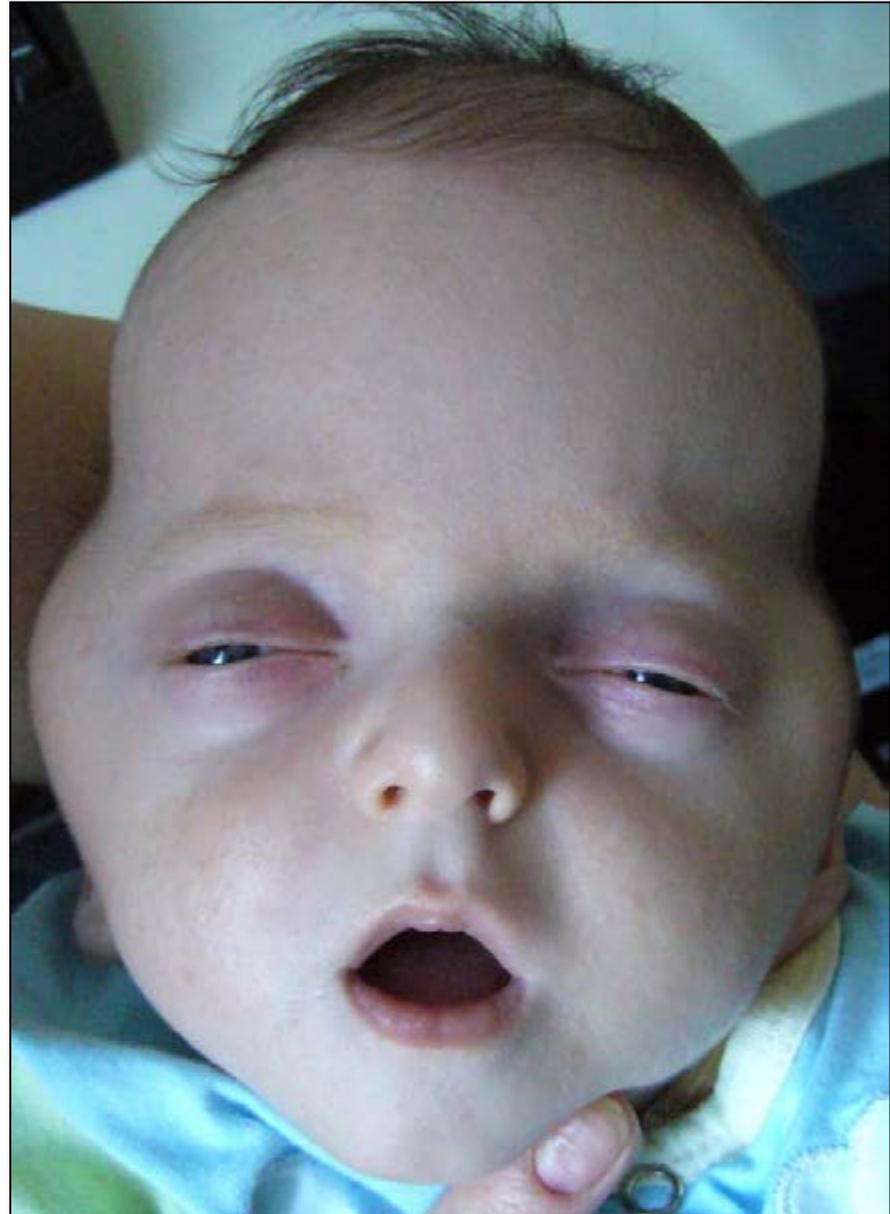


Kleeblattschädel



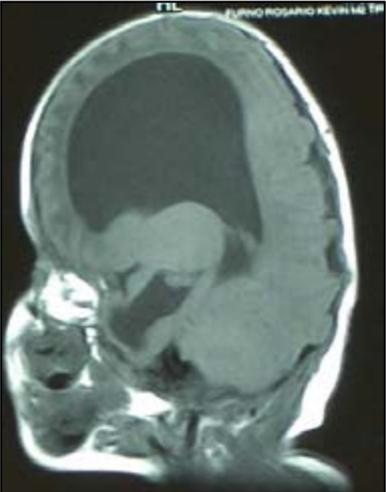
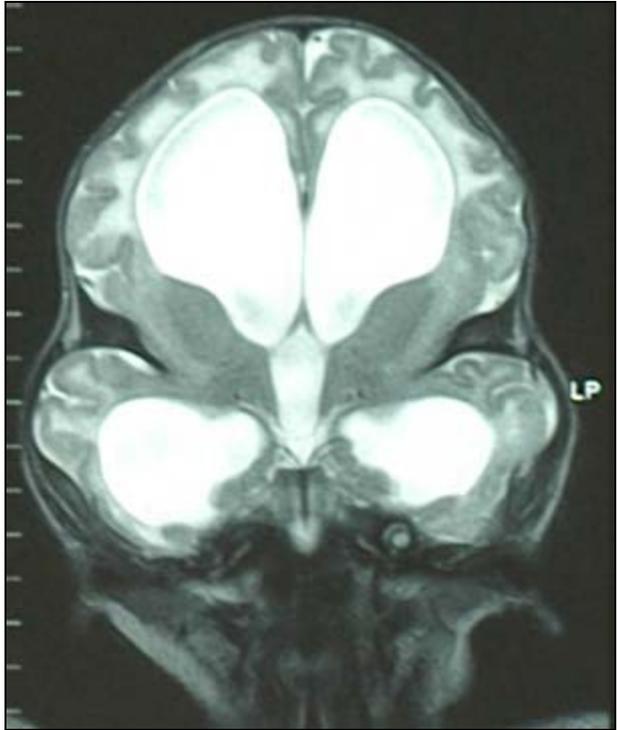
**Interessamento
suturario multiplo**

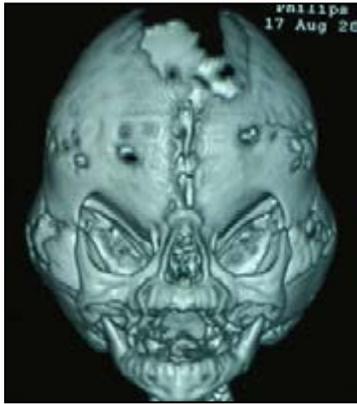
Difficili da classificare





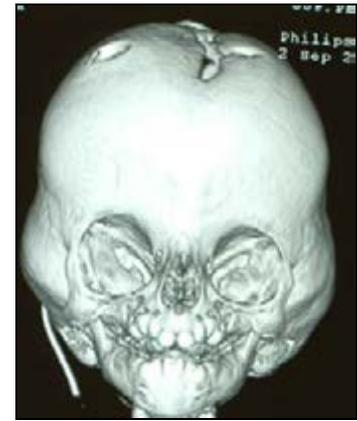
Idrocefalia
Chiari severo





PRE 3D-CT

AFTER 13 MONTHS

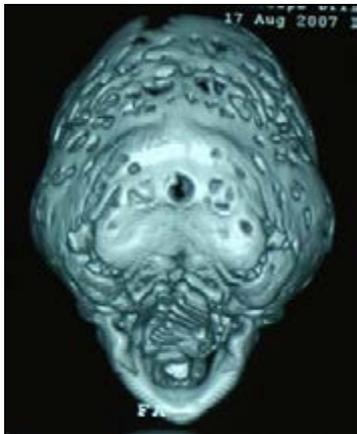
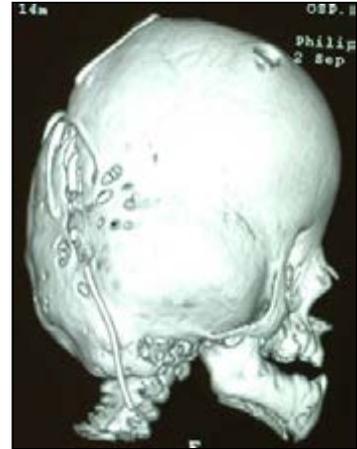


SURGICAL STRATEGY

1/ ETV

2/ SAS

3/VP shunt



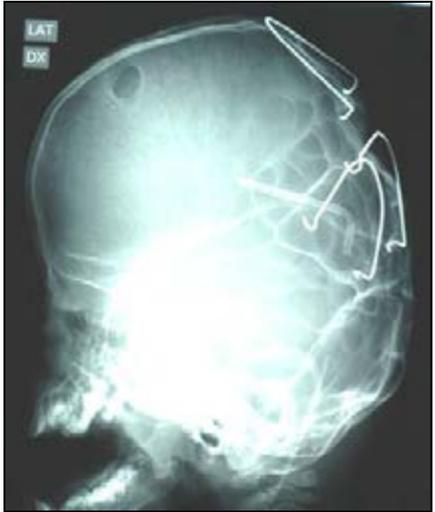
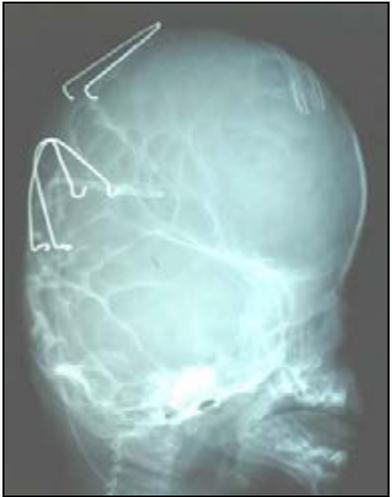
1ST POST SURGERY
CRANIAL RX

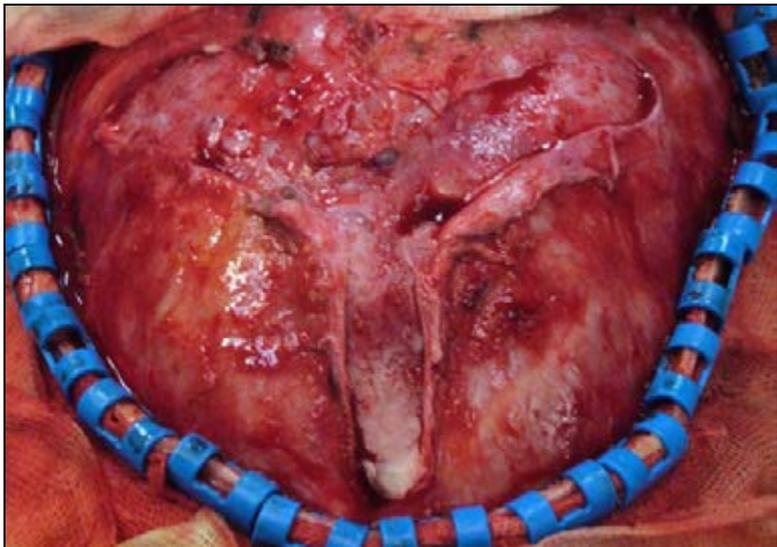
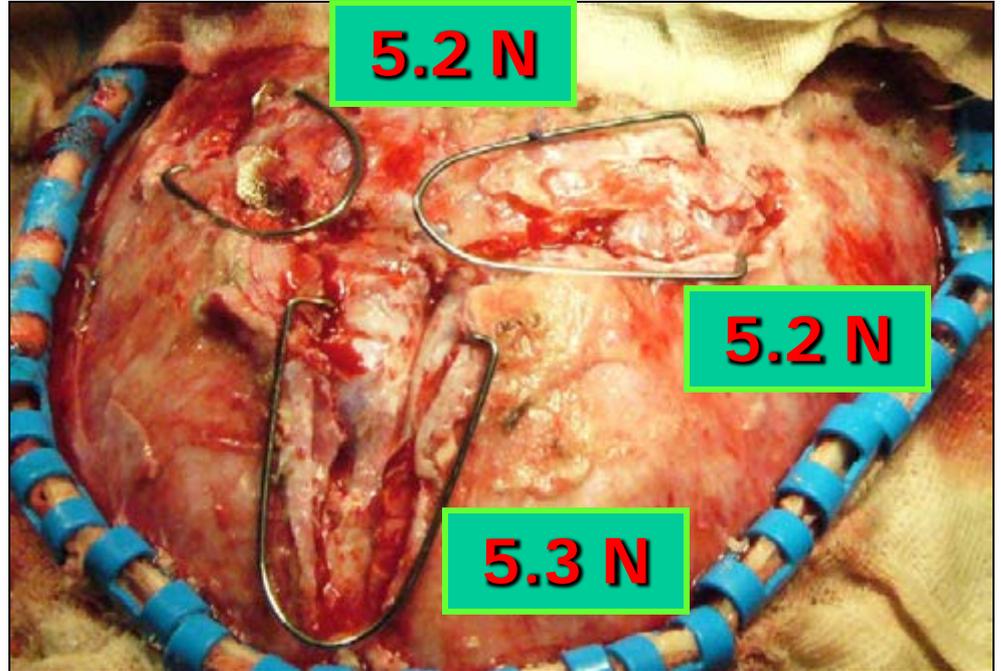


AT 2 MONTHS

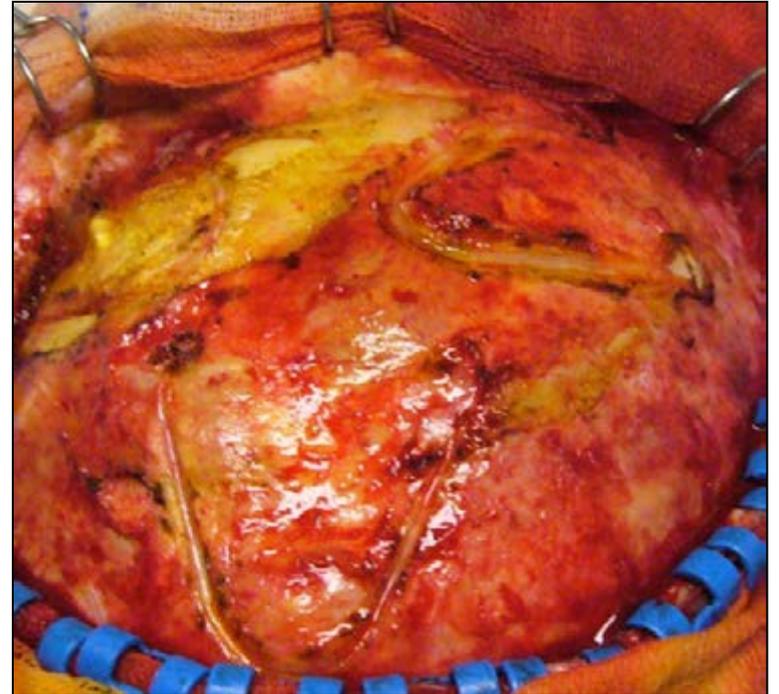
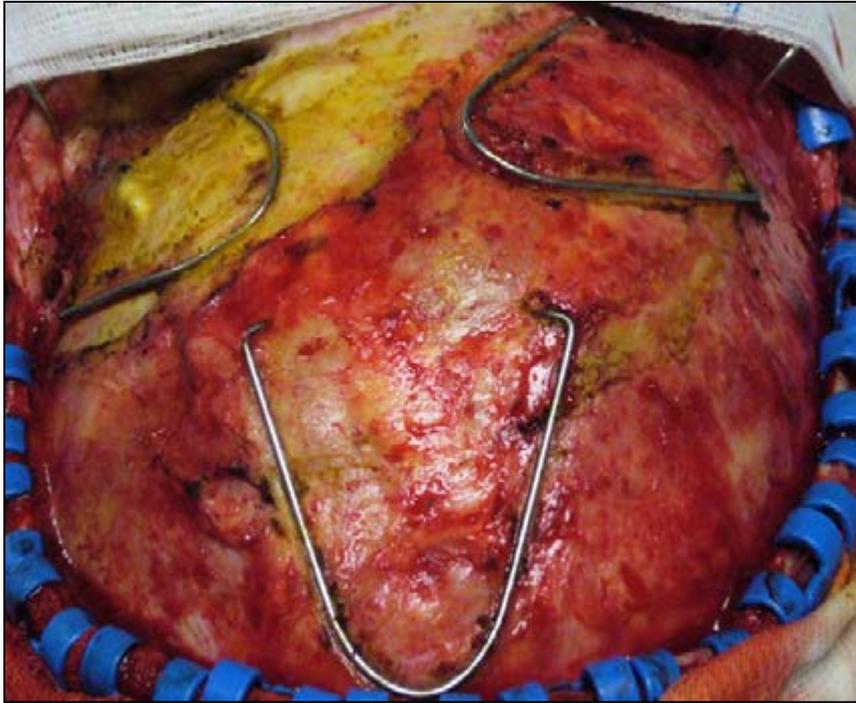


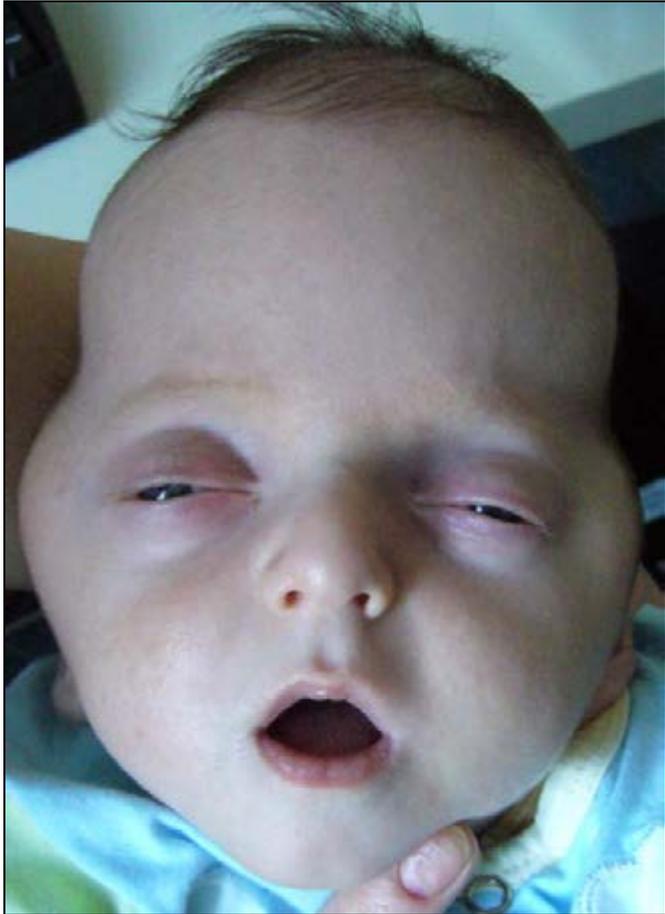
AT 13 MONTHS





REMOVAL





Conclusioni I

1. Diagnosi clinica
2. Ruolo essenziale della genetica
3. Indicazione chirurgica (età, problemi oftalmologici, estetica, ipertensione endocranica)
4. I bambini con ritardo mentale e sindromi cranio facciali presentano alterazioni cromosomiche spesso evidenti con l'analisi FISH subtelomerica
5. Impatto genetico sul risultato chirurgico: forme asimmetriche con mutazione in FGFR3 (pro250arg) e TWIST con risultati chirurgici peggiori
6. Calibrazione della tecnica e del timing chirurgico
7. RM encefalo come unico esame radiologico in casi selezionati

SPRINGS RIASSORBIBILI

Acidi polilattico e poliglicolico

(completo riassorbimento in 18 mesi



Work in progress dal gennaio 2009.....

36 casi attualmente in FU

Applicare lo stesso principio del
“distrattore”/”distanziatore” metallico
(ottenendo quindi una espansione
focale del cranio).....



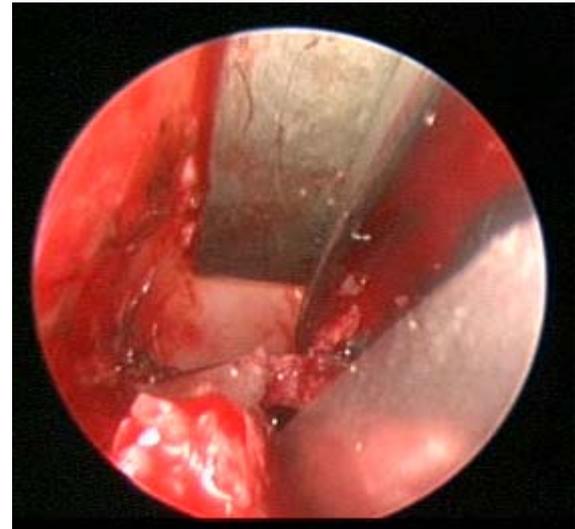
.....creando “distrattori” riassorbibili, evitando così
la necessità di secondo intervento

Tecnica mini-invasiva
68 pazienti (38 M/20F):

- 8 Scafocefalie,
 - 27 Plagiocefalie anteriori,
 - 15 Plagiocefalie posteriori,
 - 6 Pachicefalie,
 - 7 Trigonocefalie
-
- 1 Oxicefalia,
 - 2 Brachicefalia,
 - 2 Sindromi di Pfeiffer,
 - 1 Sindrome di Apert,
 - 2 craniostenosi multisuturarie non classificabili

TECNICA MINIINVASIVA

- SUTURECTOMIA A CIELO APERTO
 - POSIZIONAMENTO ESPANSORI
 - 30 metalliche
 - 36 riassorbibili
- SUTURECTOMIA ENDOSCOPICA
 - 3 endoscopiche (in una posizionata spring riassorbibile- già inclusa nelle riassorbibili)



- età media del posizionamento: 9.7 mesi (range 1-31 mesi).
- ospedalizzazione media: 5.8 giorni (range 2-12)
- timing chirurgico medio: 53' (range 35'-135')

PITFALL

“spring metalliche”

- reintervento chirurgico conseguentemente al dislocamento delle springs metalliche accorso in 3 pazienti diversi affetti da 3 differenti patologie: Sindrome di Pfeiffer, pachicefalia e plagiocefalia anteriore.



- pitfall → posizionamento dell'uncino della molla metallica sulla sutura

VANTAGGI

- riduzione della perdite ematiche (passando da 190 cc a 20 cc)
- riduzione della necessità di trasfusione
- minore analgesia post-operatoria,
- riduzione dei giorni di ospedalizzazione
- Riduzione dei costi per singolo paziente.
- riduzione dei tempi chirurgici di quasi 3 volte passando da una media di 156 minuti ad una di 53 minuti.

RISULTATI

- ECCELLENTE (80%): simmetria craniofacciale, normalizzazione della forma del cranio, dimensioni e risultati estetici piacevoli al team
- BUONO (13.4%): risultati estetici accettabili dalla maggior parte ma decretati non ideali. Generalmente secondari ad una lieve asimmetria
- INSUFFICIENTE (5.7%): presenza di una asimmetria evidente o una forma/dimensione cranica non accettabile

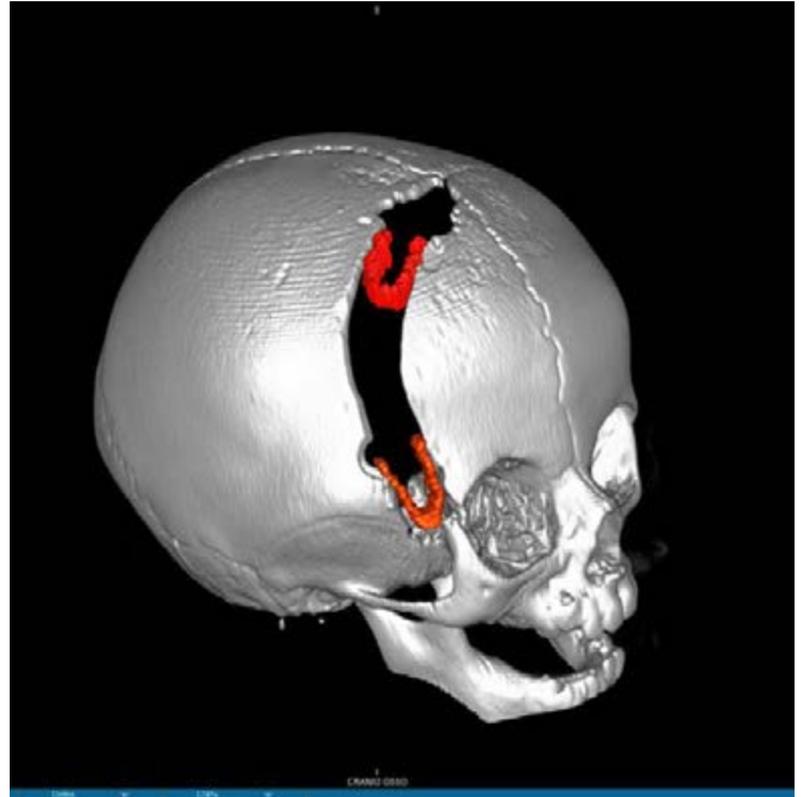
Fenotipo Pfeiffer con lussazione dei globi oculari



Plagio ant dx.



Pre op



TAC 3D

Craniotomia e
posizionamento spring
riassorbibili

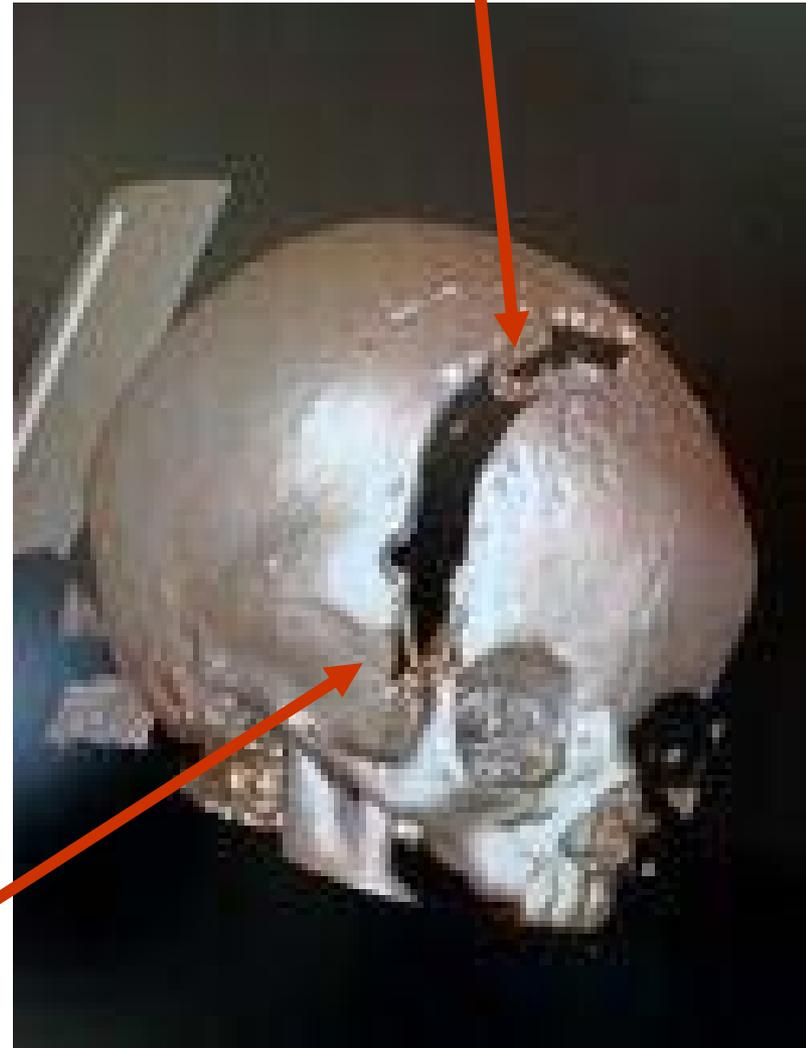
Durata intervento: 45min

Non trasfusione ematica

Durata ospedalizzazione : 3gg

Spring inf.

Spring sup.

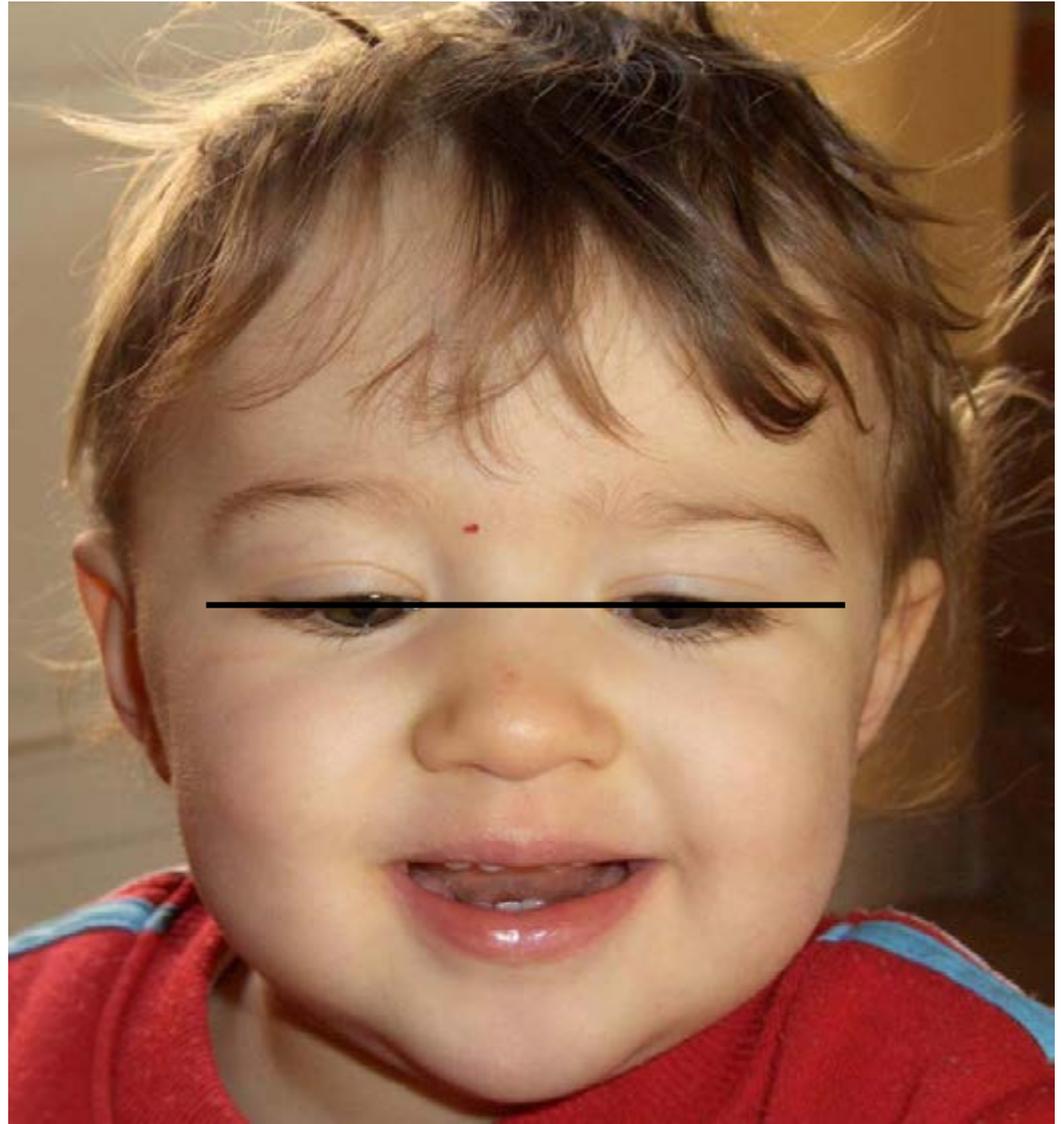


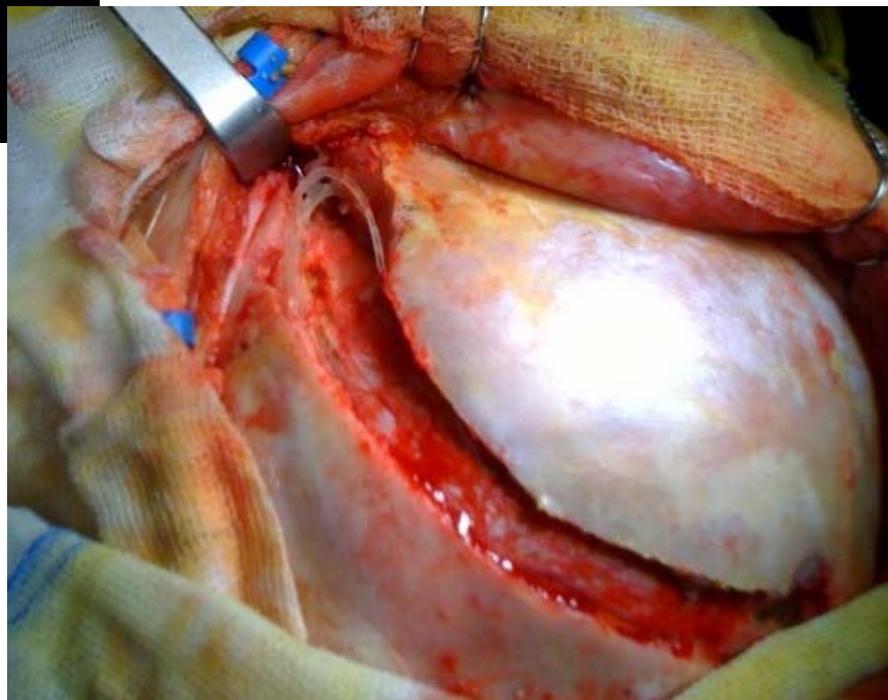
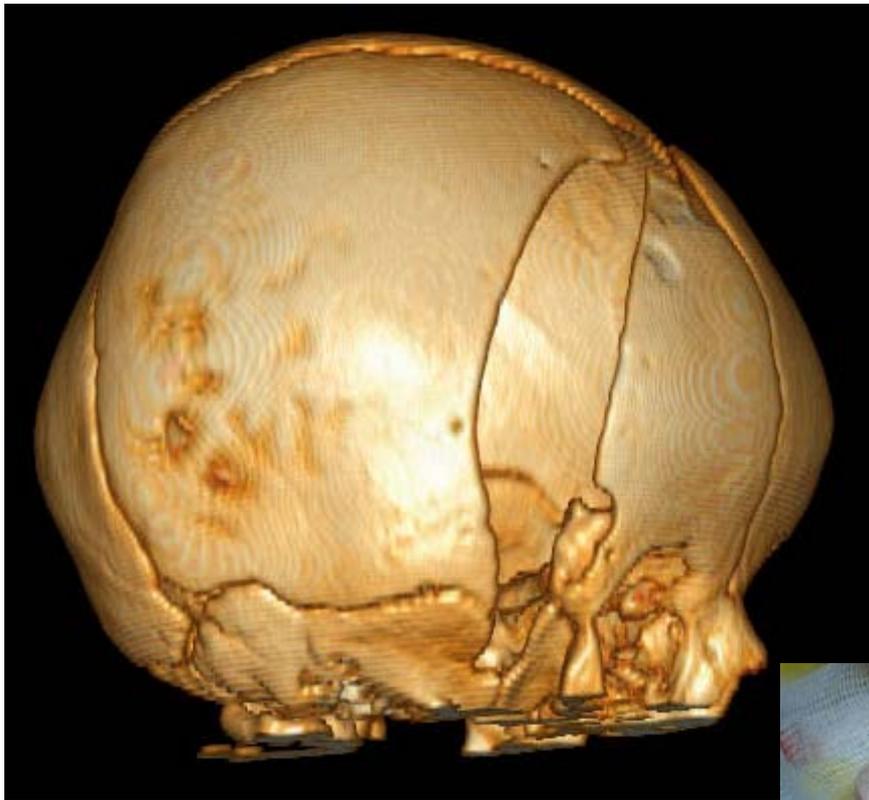
RISULTATI

Dopo 5 mesi



PRE-OP

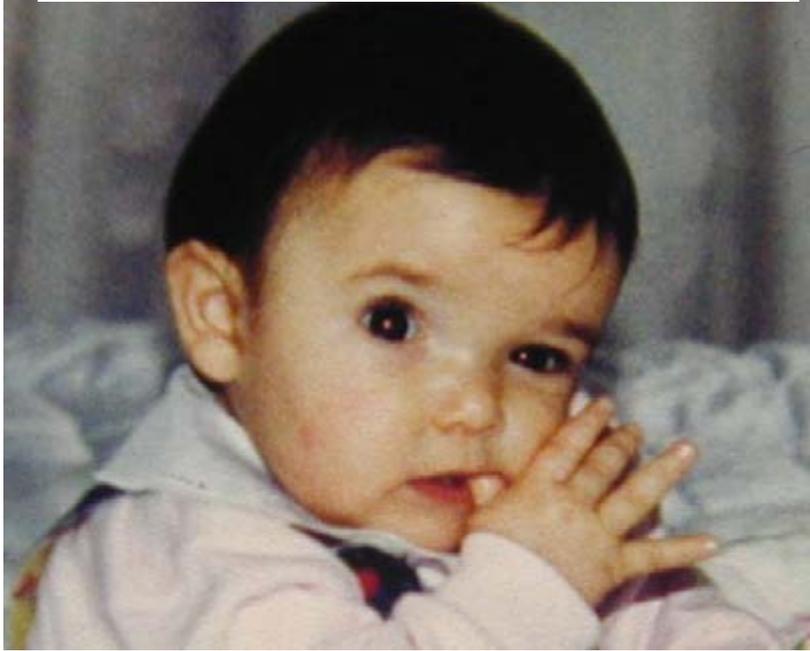




Risultati



Plagiocefalia anteriore

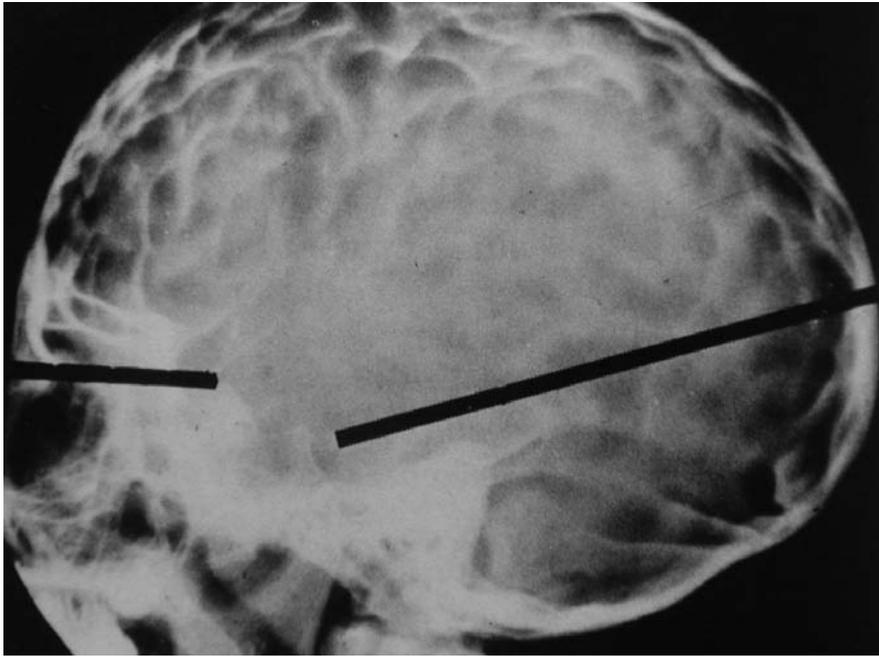


Pre op

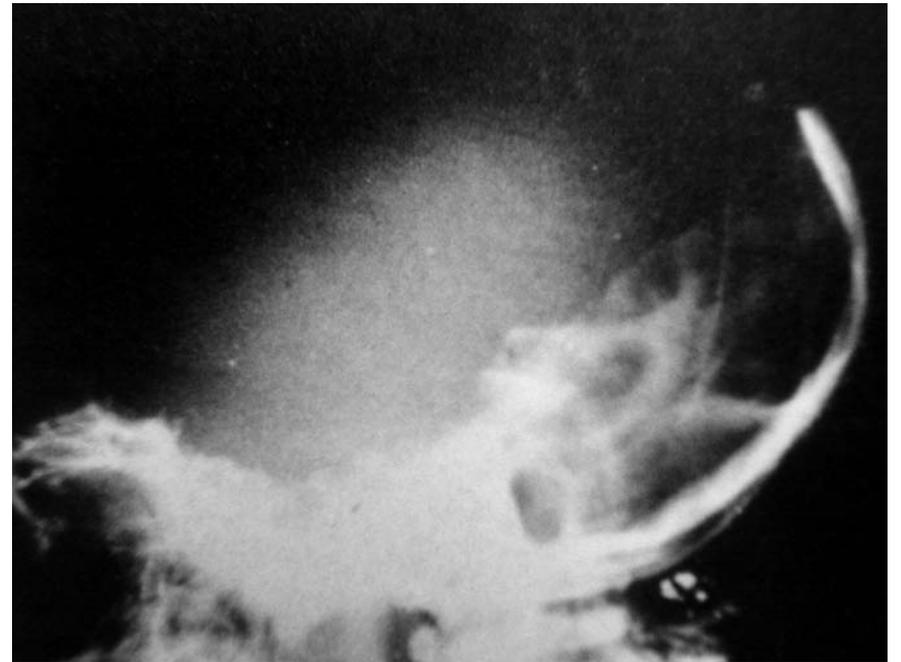
Risultati



Dopo 12 mesi



1980: Calvariectomia
completa per il trattamento
di una Craniosinostosi



CONCLUSIONI II

- 
- 1/ La Spring Assisted Surgery offre in casi selezionati una eccellente espansione di aree selezionate del cranio
 - 2/ Permette di espandere allo stesso tempo aree diverse del cranio
 - 3/ Nei casi piu' complessi e' raccomandabile una chirurgia mini-invasiva in piu' steps
 - 4/ Nelle craniostenosi piu' semplici e' raccomandabile l'uso della chirurgia mini-invasiva con spring riassorbibili



GRAZIE!