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Management of non-syndromic craniosynostoses in France in 2015: A national survey

Raphaëlle Prevost [a, *], Pierre Keribin [c], Claire Batut [d], Benjamin Guichard [d], Béatrice Ambroise [b], Alin Bohra [e], Hervé Benateau [b], Alexis Veyssiere [b]

[a] Department of Oral and Maxillofacial Surgery, La Rochelle Hospital, 17000 La Rochelle, France
[b] Department of Plastic and Oral and Maxillofacial Surgery, Caen University Hospital, Avenue de la Côte-de-Nacre, 14000 Caen, France
[c] Rouen University Hospital, Department of Oral Surgery and Odontology, F 76000, France
[d] Rouen University Hospital, Department of Oral and Maxillofacial Surgery, F 76000, France
[e] Neurosurgery department, Caen University Hospital, Avenue de la Côte-de-Nacre, 14000 Caen, France

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ABSTRACT

Purpose: Craniosynostoses are managed by surgical and anaesthetic teams in specialist centres. Despite the availability of international guidelines, the perioperative management of craniosynostoses remains highly variable between centres. The aim of our study was to describe the different protocols for the management of non-syndromic craniosynostoses in France in 2015.
Materials and methods: This descriptive study consisted of a survey about the protocols of pre-operative, per-operative and post-operative management of craniosynostoses. The survey was sent to the departments in French university hospitals that perform this surgery.
Results: Nineteen departments out of twenty replied to the survey. Sixteen departments organised multi-disciplinary meetings. The most frequent preoperative imaging requested was a Computed Tomography. More than half of the centres organised a follow-up until early adulthood.
Conclusion: This study showed a great variability in the management of craniosynostoses. A recommendation from the study is to establish a scientific committee of practitioners in order to establish a standardised protocol. In addition, this study showed the need to create a specific section in the French rare diseases database (CeMaRa) for craniosynostoses.

1. Introduction

Non-syndromic craniosynostoses affect 1 out of 2000 newborns in France (Renier et al., 2006a). There is no consensus in the definition of non-syndromic craniosynostoses due to the variety in presentation.

They are managed in specialised centres by dedicated surgical and anaesthetic teams. Surgical correction is not recommended for mild types of craniosynostoses such as metopic ridge and partial synostosis of the sagittal suture with little morphologic impact. For all other types, surgery aims to correct the anomaly for functional and morphological reasons (Mathijssen, 2015). In France, there is a consensus to perform the surgery before the age of 1 year. This allows a harmonious restoration of the morphology and limits the risks of severe functional complications. Further, intracranial hypertension increases with age and is an important risk factor for neurological and ophthalmological disorders (Marchac and Renier, 1989; Renier et al., 2006b, 2006c).

However, several parts of this highly specialised management are still controversial. Most centres organise a multi-disciplinary management (Birgfeld et al., 2015). However, in other centres, a single surgeon decides the entire plan with no consultation.

The techniques of reconstruction are more complex for major deformations and as patients grow older. In addition, there is also a great diversity in the surgical techniques and no consensus regarding the peri-operative management, which varies from one centre to another.

* Corresponding author.
E-mail addresses: raphaelle.prevost@gmail.com (R. Prevost), pierre.keribin@gmail.com (P. Keribin), claire.batut@gmail.com (C. Batut), guichard@cmfrouen.fr (B. Guichard), ambroise-b@chu-caen.fr (B. Ambroise), borha-a@chu-caen.fr (A. Bohra), benateau-h@chu-caen.fr (H. Benateau), veyssiere-a@chu-caen.fr (A. Veyssiere).

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The aim of this study was to describe the medical and surgical protocols used for the management of non-syndromic craniosynostoses (scaphocephaly, anterior plagiocephaly and trigonocephaly) in France in 2015.

2. Materials and methods

This descriptive study explored professional practices in the management of craniosynostoses. Even though there is no organised network for the management of craniosynostoses, only university hospitals have the infrastructure to treat these rare congenital disorders. All departments of Cranio-Maxillofacial Surgery and Neurosurgery in these university hospitals were contacted. A survey with 79 questions was then sent to the departments that confirmed them to be involved in the management of craniosynostoses. The survey considered only non-syndromic craniosynostoses. "Non-syndromic" was defined in the survey as isolated craniosynostoses, without any other morphological or neurological anomaly clinically found.

The survey focused on the composition of the teams, the initial review, the anaesthetic management, the age of surgery, the peri-operative management, the post-operative surveillance, and the long-term follow-up.

One response per centre was analysed, as it was expected that practices were similar inside each team, regardless of the specialty of the consultant surgeon who returned the survey.

3. Results

A total of 20 university hospitals out of 32 confirmed managing craniosynostoses. Of these, 19 returned the survey: Angers, Amiens, Besançon, Bordeaux, Caen, Clermont-Ferrand, Grenoble, Lyon Croix-Rousse and Lyon Pierre Wertheimer, Marseille, Montpellier, Nancy, Nice, Paris (Hôpital Necker-enfants malades), Rennes, Rouen, Toulouse, Tours and Strasbourg.

Only consultant surgeons answered and returned the survey. Results are presented in Tables 1–5.

4. Discussion

This is the largest survey on the peri-operative management of craniosynostoses in France. The results reveal major differences in the treatment between the 19 French university hospitals. At the time of the study, there was no national network organised for the management and the collection of data for this rare congenital disorder. However, some of these centres already belong to the European Reference Network on Craniofacial Anomalies and ENT Disorders. This study aimed to emphasize the lack of standardisation in the management of craniosynostoses and the need to create a national network.

4.1. Composition of the teams and initial assessment

This study shows a large variation in the composition of the teams. Three were composed of a single practitioner, 14 with surgeons only and 5 teams included a medical practitioner.

Studies have shown that multi-disciplinary teams improve the quality of initial assessment and further management (Dhellemmes et al., 2002; Stricker, 2003; Meyer et al., 2006; Arnaud et al., 2006; Birgfeld et al., 2015). It is recommended to include at least 1 Neurosurgeon (Mathijssen and Arnaud, 2007) and 1 Facial Surgeon, focusing on functional and aesthetic outcomes and safety. Additionally, this team should include a Geneticist or Paediatrician to detect genetic syndromes and evaluate the psychomotor development (Mathijssen, 2015; McCarthy et al., 2012). Joint consultations should be held and teams should convene on a regular basis (American Cleft Palate–Craniofacial Association, 1993). Non-core specialties do not need to attend every consultation but should be available if assessment is required (Capone and Sykes, 2007).

Finally, the initial assessment should include photographs, a psychomotor development evaluation and an ophthalmic examination with ophthalmoscopy (Marchac and Renier, 1989; Strohecker, 1993). This is essential for early detection and management of the causes of impaired vision such as optic atrophy, corneal abnormalities in lagophtalmos or amblyopia secondarily to strabismus or refractive errors (Mathijssen, 2015).

4.2. Imaging

Risks from imaging examinations include radiation exposure. This is a controversial topic due to the consequences of these radiations. The estimated increased risk of cancer over a patient's lifetime from a single CT scan is limited (estimated to be 0.03%–0.05%) (Mathijssen, 2015). However, the consensus is to limit radiation exposure.

4.2.1. Pre-operative imaging

Different examinations are routinely used by the teams. Renier et al. and Lupescu et al. recommend in their studies the use of a CT with three-dimensional (3D) reconstructions (Lupescu et al., 2000; Renier et al., 2006d; McCarthy et al., 2012) even if the diagnostic of craniosynostoses is first based on clinical information.

Table 1
Pre-operative assessment

Pre-operative assesment						
In your hospital, to which specialist are patients presenting with craniosynostoses referred? Is there a multidiscliplary team? If so, which specialists are involved in this team?						
Teams	Multi-disciplinary	Single practitioner				
	16 (85%)	3 (15%)				
Specialties	Neurosurgeon	Maxillofacial Surgeon	Plastic surgeon			
	19 (100%)	12 (63%)	6 (31%)			
Other members	Geneticist	Pediatrician	Nurse	Psychologist	Photographer	
	3 (15%)	2 (10%)	2 (10%)	2 (10%)	1 (5%)	
Which pre-operative imaging examinations are performed for scaphocephaly, plagiocephaly and trigonocephaly?						
	None	X-ray	CT-scan ± 3D	X-ray + Ct-scan	CT + MRI	MRI
Scaphocephaly	2 (10%)	1 (5%)	10 (53%)	5 (26%)	1 (5%)	0
Plagiocephaly	0	1 (5%)	11 (58%)	5 (26%)	2 (10%)	0
Trigonocephaly	0	2 (10%)	9 (48%)	5 (26%)	2 (10%)	1 (5%)
What do you use in your pre-operative anaesthetic protocol to prepare the patient for surgery?						
	Iron supplements	Erythropoietin injection	None			
	2 (10%)	10 (53%)	7 (37%)			

Table 2
Surgical management.

Surgical management						
What is the ideal age considered for surgery for scaphocephaly, plagiocephaly and trigonocephaly?						
	2–4 months	3–6	6	6–9	9	9–12
Scaphocephaly	7 (37%)	10 (53%)	0	2 (10%)	0	0
Plagiocephaly	0	0	4 (21%)	6 (31%)	4 (21%)	5 (26%)
Trigonocephaly	0	0	4 (21%)	5 (26%)	4 (21%)	6 (31%)
What shape of incision do you routinely use?						
	Broken line	Coronal	Other			
	13 (68%)	4 (21%)	6 (31%)			
Do you use drains? If yes, how many?						
	None	1	2			
	13 (68%)	4 (21%)	2 (10%)			
What do you use for skin closure?						
	Resorbable sutures	Non-resorbable sutures	Staples			
	17 (90%)	1 (5%)	1 (5%)			

Table 3
Post-operative period.

Post-operative period						
With no complication, are the patients staying in the intensive care unit? Overall, what is the average length of stay in the hospital?						
ICU	Yes (24–48 h)	No				
	17 (90%)	2 (10%)				
Length of stay	4 days	5	6	7	8	9
	1 (5%)	6 (31%)	4 (21%)	3 (15%)	4 (21%)	1 (5%)
Which post-operative imaging examination do you routinely ask for?						
	None	CT-scan	Frontal X-rays	X-rays + transcranial ultrasound		
	12 (63%)	3 (15%)	3 (15%)	1 (5%)		
Do you prescribe an antibiotic treatment? If Yes, for how long?						
	None	Per-operative only	1 day	2	5	10
	7 (37%)	7 (37%)	2 (10%)	1 (5%)	1 (5%)	1 (5%)
Which wound care do you prescribe for discharge?						
	Nurse wound care	Antiseptics	Empirical antibiotic treatment			
	2 (10%)	3 (15%)	1 (5%)			

Table 4
Long-term follow-up.

Long-term follow-up						
How long after the surgery do you plan your first follow-up appointment?						
1 week	2	1 month	1.5	2	3	6
2 (10%)	1 (5%)	4 (21%)	2 (10%)	1 (5%)	8 (43%)	1 (5%)
At what age do you end the follow-up?						
5–6 years old	7–8	12	15–18	20		
4	1 (5%)	2 (10%)	11 (58%)	1 (5%)		
Is the follow-up completed by a multi-disciplinary team?						
	Multi-disciplinary team	Single practitioner				
	11 (58%)	8 (43%)				
Which long-term imaging examination do you routinely ask for?						
	None	X-rays	CT-scan	X-rays and/or CT-scan		
	6 (32%)	5 (26%)	4 (21%)	4 (21%)		
Do you routinely organise an ophthalmologic follow-up? A neurological assessment?						
	Yes	No				
Ophthalmologic	11 (58%)	8 (43%)				
Neurological	0	19 (100%)				

The MRI is not systematic and appears to be more appropriate for genetic syndromes or abnormal cerebral development (Dhellemmes et al., 2002).

4.2.2. Post-operative imaging

The responses from the survey for post-operative imaging examinations varied among the centres with only 3 requesting a CT scan. Considering the risk associated with the radiation exposure, McCarthy et al. questioned the need for routine postoperative CT scans. These CT scans are done to eliminate complications such as hematoma or ischemia. However, the centres that do not use post-operative CT scans do not appear to have more complications. In the post-operative period, any complication must be detected by a clinical examination and CT scans must be done only to confirm the diagnosis and not routinely (Renier et al., 2006d).

4.3. Age of surgery

In France, it is common to prioritise early surgery to limit the risks of neurological complications (Renier et al., 2006b; Patel et al., 2014), while also considering the anaesthetic risks (Meyer et al., 2006; White et al., 2015).

A conventional surgery should be performed before 6 months for scaphocephalies, and between 6 and 9 months for other malformations (Marchac and Renier, 1989; Arnaud et al., 2006).

Table 5
Complications.

Complications	
How many complications among the below items occurred in the last 3 years?	
None	6
Soft tissue infection	4
Exposure of surgical material	3
Skin necrosis	4
Diplopia	1
Hemorrhage	1
Chronic subdural hematoma	1
Cerebral emphysema	0
Post-operative meningitis	0
Death	1

Anaesthetists have a crucial role in managing young patients undergoing a major procedure and should be involved in treatment decisions. Studies highlight the benefits of using tranexamic acid to limit blood loss (Dadure et al., 2011; Martin et al., 2015).

4.4. Choice of incision

Coronal incisions are used for most comprehensive procedures. The area of most visible scarring is typically the temporal region because the scar is in line with the orientation of the hair follicles. Posteriorly inclined incision lines are less visible because of oblique and perpendicular folliculi orientation adjacent to the scar. Zigzag modifications of the incision line in the temporal region have proven to enhance this effect (Persing, 2008). In the current survey, these aesthetic considerations may explain why 68% of the surgeons reported using this pattern.

4.5. Length of stay in hospital

Recent guidelines recommend a post-operative stay in intensive care unit to monitor blood loss and allow better pain management (Mathijssen, 2015). In our study, 90% of centres routinely admitted their patients for 24–48 h after surgery.

The average total length of stay in hospital in the absence of complications was between 4 and 9 days. This appears to be longer than in previous studies (Alperovich et al., 2015). In the current study, the post-operative wound care and dressings were completed during the hospital stay. Patients were discharged without any further care in 14 centres.

4.6. Antibiotic treatment

The risk of infection in craniosynostosis surgery is extremely low when not associated with entering the nasopharynx. In addition, prolonged use of antibiotics is related to the possibility of allergic reactions and selection of resistant bacteria (Persing, 2008).

In the current study, all centres reported no severe post-operative infections. The low rate of infection may be related to the good vascularisation of the scalp skin and to the young age of the patients (Persing, 2008). Seven centres were not prescribing any prophylactic antibiotic treatment, seven centres were limiting their use during the peri-operative period and 5 centres were prescribing a prophylactic post-operative antibiotic for 1–10 days (Table 3).

Currently, there are no recommendations about the use of antibiotics for craniosynostoses surgery. However, Hunter demonstrated the absence of benefit to start a prophylactic antibiotic treatment after wound closure and that it should not be continued for more than 24 h after the end of surgery (Hunter, 2007). Considering the risks reported above, the low rate of infection and the absence of benefits reported, these prolonged post-operative treatments appear to be questionable.

On the other hand, the French Society of Anaesthesia recommends a perioperative antibiotic treatment for craniotomies performed on adults (Société Française d'Anesthésie et Société française d'anesthésie et de réanimation, 2011). In the absence of consensual paediatric recommendations, these guidelines could be adapted for craniosynostoses surgery. They consist of one injection of first-generation cephalosporin (or vancomycin if the patient is allergic) every 4 h during surgery (Société Française d'Anesthésie et Société française d'anesthésie et de réanimation, 2011).

Further studies dedicated to the use of antibiotics for craniosynostoses would be necessary to establish clear recommendations about the need and the ideal length of these treatments.

4.7. Surgical complications

The surgeons were asked to report their complications in the last 3 years. Medical records were not verified. The number of complications in the current survey was low, and superficial infections represented the majority. One centre reported one fatal case, coherent with the incidence of less than 1% described in the literature (Fearon et al., 2009). Another complication is related to the use of wires that may become exposed. To overcome this, the use of resorbable material should be considered. As the question focused on the last 3 years, bone defects and long-term aesthetic outcomes were not studied.

4.8. Follow-up

A surgical follow-up should be organised in the first 3 months, then annually until the end of adolescence. This would allow photographic assessment of long-term morphological results (Sainthillier et al., 2009).

In addition to these morphological results, functional outcomes should also be considered. An ophthalmologic follow-up is commonly recommended with 11 centres reporting organising this follow-up routinely. On the other hand, neuropsychological follow-up is not included in the 19 centres' protocols. The impact of craniosynostosis surgery on neuropsychological development is also poorly documented in the literature. It would be interesting to determine how cranio-facial deformations and surgical procedures on young patients' skulls can affect their development. Adding a neuropsychological follow-up would allow the collection of data for future studies to establish whether or not it would be beneficial for patients in routine practice.

4.9. Study limitations and strengths

A limitation of this study was that the survey did not ask how many craniosynostosis surgeries were performed annually. This could have exposed surgeons who manage a low number of cases every year to have their competency and their ability to perform craniosynostosis surgery in the future questioned. As a result, this question was not included in the survey so as to maximize the number of responses. In addition, in the absence of national data for craniosynostoses, no official statistics on the annual number of surgeries per centre were available.

This study showed the need to organise a national scientific committee to establish a protocol for the management of craniosynostoses. Furthermore, this study showed the importance of having a national network and database. As this is a rare condition, these data would assist future decision making about the centralisation of the management of craniosynostosis surgeries.

In the absence of standards, analyses are impossible, particularly given that in some teams, follow-up ends around age 5–6 years, which prevents the collection of long-term data. This protocol would standardise practice, would benefit patient and would allow for evidenced based multi-centre studies, comparable to the major studies conducted in the United States. These US studies are based on data collected through a national database specifically designed for major paediatric surgical procedures (Czerwinski et al., 2010; Chow et al., 2015). CeMaRa (Centre for Rare Diseases) is the French equivalent database. It elaborates and evaluates strategies to improve the management of rare diseases by collecting a limited amount of information for every patient, according to that patient's pathology. The creation of a section in the CeMaRA database focusing on non-syndromic craniosynostoses would help the teams to collect clinical information and photographs.

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