Cleft-Palate And Hare-Lip: The Earlier Operation On The Palate (Classic Reprint)

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Anatomy for Plastic Surgery of the Face, Head, and Neck details the complex regional anatomy of the face, head and neck, providing plastic surgery and otolaryngology residents with a solid anatomical knowledge base.

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Second Edition: First Printing. Boards and pages are clean, unmarked, brightly colored, tightly bound and sharp cornered but has slight scuffing at top of spine from shelf wear.

Book is in English, translated from the original German. Contains illustrations. Scarce, Out Of Print, Book. This book is issued as part of a three
In , he advanced palatal surgery even further with the introduction of relaxing incisions to ease palatal closure. After his death in , Dieffenbach was director of the Clinical Institute of Surgery at Charite Hospital in Berlin, first described closure of the hard palate by mucosal elevation.

The technique was good for speech. Postoperative palatal fistulas were encountered in four patients in the study. In , Johann Friedrich Dieffenbach, the many patients showed improvement in their 6-month postoperative speech in both groups. The preoperative values of speech assessment all three complications occurred throughout the study except in one patient who had secondary bleeding after 48 hours of the surgery. There were 12 males and 8 females. The age at repair ranged from 18 months to 36 months mean age.

Intelligibility was graded into- C1 intelligible at all times, C2 sometimes unintelligible, C3 unintelligible most of the time. Velopharyngeal speech or three or more errors but intelligible, B4 multiple errors, frequently unintelligible. B1 normal, B2 one to two consistent errors only with no deterioration in speech, B3 one to two consistent error with deterioration in connected articulation problem or fluid regurgitation from the nose.

Speech was classified into three different groups, namely A nasality, B articulation, C intelligibility, each of which is further subdivided by numbers. Nasality was divided into A1 normal, A2 mild hyper nasality, A3 moderate hyper nasality, A4 severe hyper nasality. Articulation was divided into B1 normal, B2 one to two consistent errors only with no deterioration in speech, B3 one to two consistent error with deterioration in connected speech or three or more errors but intelligible, B4 multiple errors, frequently unintelligible.

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All the children underwent speech assessment and the counseling. In all the patients following variable were determined: quality of speech, incidence of fistula, and age at the time of surgery. Clinically significant fistulas were determined by the presence of either hypernasal speech, articulation problem or fluid regurgitation from the nose.

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occur with cognitive impairments and behavioral disorders. This can be a source of stress for parents and lead to a delay in diagnosis and treatment. Nonsyndromic and syndromic craniosynostosis may co-

professionals who do not recognize the craniosynostosis shortly after the birth of the child.

Deformities of the extremities are notably restricted to the syndromic craniosynostoses, and in patients with Apert syndrome always present in a severe form. In the other syndromic types, deformities of the extremities are generally very mild. Parents are often confronted with health

reviews.

Abstract Treatment of the cleft palate has evolved over a long period of time. Keywords: Cleft palate, palatoplasty, speech assessment.

Palatoplasty technique Twenty patients in the age group of 18—36 months underwent Pinto's modification of Wardill—Kilner two-layer palatoplasty without radical dissection soft palate musculature. Table 1 Speech result. Open in a separate window. Table 3 Intelligibility. Figure 1.

Cleft-Palate And Hare-Lip: The Earlier Operation On The Palate (Classic Reprint) Reviews

Deformities of the extremities are notably restricted to the syndromic craniosynostoses, and in patients with Apert syndrome always present in a severe form. In the other syndromic types, deformities of the extremities are generally very mild. Parents are often confronted with health professionals who do not recognize the craniosynostosis shortly after the birth of the child.
These may occur both intrinsic to the congenital defect and secondary to, for example, increased cranial pressure or abnormal physical appearance. Notably, families of a child with syndromic craniosynostosis may experience psychosocial problems, such as having to cope with negative reactions from others, a possible discrepancy between deviating physical appearance and cognition, and problems with school choice.

How to optimize recognition of craniosynostosis in the primary and secondary healthcare sectors? Craniosynostosis should be recognized timely for optimal treatment. It appears, however, that craniosynostosis patients often are not referred at all or referred too late. A complicating factor in the recognition of craniosynostosis is the high incidence of positional head shape deformities. It may be expected that recognition, and thus referral of patients with craniosynostosis will happen earlier if health professionals in the primary and secondary healthcare sectors are facilitated to make this distinction.

Preceding the referral to a tertiary healthcare center, we often see overuse of diagnostic imaging, which is associated with further delay in referral, an extra burden and insecurity for patient and parents as well as unnecessary expenditure.

This should be restricted to a minimum. There are many options to diagnose isolated craniosynostosis or any associated abnormalities. To restrict both underdiagnosis and overdiagnosis, basic question 3 concerns further diagnostic procedures in a tertiary healthcare center. Areas of attention are diagnostic imaging, genetic diagnostics, and the role of the pediatrician.

Inclusion of a clinical geneticist in a multidisciplinary craniofacial team provides the opportunity to address questions from parents and attending physicians. For the attending physician it is important to know if there are any associated anomalies to be expected next to the craniosynostosis that may be of influence to the treatment plan and the prognosis of the child.

The abnormal skull shape is recognized by parents themselves, midwife, obstetrician, general practitioner, infant health clinic physician, nurse, pediatric physiotherapist, pediatrician, pediatric neurologist, and helmet maker.

Occasionally, it is recognized later by other specialists and sometimes not until a craniofacial team is involved. These data are from the Birmingham craniofacial center and could therefore well compare with the situation in the Netherlands.

Of the 89 referrals with the initial diagnosis nonsynostotic occipital plagiocephaly NSOP made by the referrer, 10 patients appeared to have a craniosynostosis. A craniosynostosis or positional skull deformity is primarily recognized by physical examination, notably skull shape in combination with history taking, and an imaging study is rarely indicated.

This deserves great attention because early recognition and surgical correction are essential to a satisfactory treatment outcome. Ridgeway recommends a number of diagnostic steps that should lead to early diagnosis, in which history taking and physical examination yield the most important information to distinguish between positional head shape deformities and craniosynostosis.

Using this information in a flowchart at intake appears to be a safe method to make this distinction at the earliest. This will prevent delay in start of treatment as a possible consequence of the increase of patients of positional head shape deformities.

There are no other publications on referral patterns in children with abnormal skull shape. History taking should address the distinction between positional head shape deformities and craniosynostosis on the one hand, and risk factors and associated anomalies on the other hand.

Craniosynostosis is present at birth, positional head shape deformities usually not. Presence or absence of a preferred sleep position is essential in the diagnosis of positional head shape deformities.

Analysis of a flowchart to differentiate positional head shape deformity versus craniosynostosis by means of a questionnaire completed by telephone or e-mail showed that the questionnaire recognized no more than More in detail the different craniosynostoses can be recognized as follows. The anterior fontanel shows earlier fusion or is triangular shaped rather than diamond shaped in synostosis of 1 adjacent suture. Mid-face hypoplasia, proptosis, and hypertelorism are found particularly in the syndromic types of craniosynostosis.

The following characteristics may help to distinguish craniosynostosis from positional head shape deformities: American Cleft Palate-Craniofacial Association ACPA recommends that craniofacial teams provide education to health professionals in obstetrics and neonatology as well as GPs and infant health clinic physicians so as to improve early recognition of children with craniofacial deformities. Grof emphasizes that effective implementation of guidelines requires behavioral changes in physicians.

E-mail consultations improve the quality of decision-making, notably as they are easily accessible. Audit programs are most effective when integrated with other educational and research programs. Often it takes long before parents and the child with craniosynostosis are referred to the tertiary center, which may lead to treatment delay and confusion, and consequently a great deal of parental stress. The flow diagram to discriminate between positional skull deformities and craniosynostosis was validated in a tertiary care setting, where it was applied by secretaries who were not specifically trained in distinguishing between these 2 conditions.

Consequently, we do not expect great differences in its reliability when applied in primary and secondary care settings. Ideally, validation for primary and secondary care settings finds place in these settings, but this does not seem to be practically feasible in view of the very low frequency of craniosynostosis compared with positional skull deformities.

Additional imaging studies to differentiate between positional skull deformities and craniosynostosis are rarely performed in tertiary centers. On clinical diagnosis of a positional skull deformity, skull x-ray or ultrasound are advised against to prevent unnecessary medical imaging with associated costs, radiation exposure, burden for patient and parents, lack of added value, and required experience in interpretation.

Consultation with a tertiary craniofacial center is indicated in suspected patients, for which submitting normal pictures front view, lateral view,
Provide structured education and training instruction to infant health center physicians, GPs, midwives and obstetricians about over skull deformities via centers of expertise with an initiating role for the tertiary centers. Provide feedback about the referral pattern on the basis of an analysis of national registry data focus on patient's age at referral to tertiary center. The craniosynostosis centers should develop a website with relevant information for the primary and secondary healthcare sectors, providing an e-mail address for online consultations.

They should stimulate inclusion of the decision tree in the ICT-systems used by GPs and infant health center physicians, as well as in training programs for GPs, infant health center physicians, midwives, pediatricians and gynecologists.

A standardly performed three-dimensional CT scan to objectivize the deformity is highly recommended for operative planning. X-skull a-p, lateral, Towne for back of the head, Tschebul for forehead is always performed in suspected craniosynostosis. If unclear, because of very young age of the patient, it is recommended to repeat X-skull after 1 or 2 months. If X-skull confirms, or does not exclude, a synostosis, a CT-scan with three-dimensional reconstruction is performed.

It appeared that ultrasound could well visualize a craniosynostosis in 26 children with craniosynostosis aged from 2 to 7 months and in 23 of the 24 children with craniosynostosis aged from 1 to 11 months. The clinical diagnosis of craniosynostosis by means of physical examination forms the basis.

Even though an X-skull is not always reliable, notably at very young age several months old, as first further diagnostic test, it provides much information and can exclude craniosynostosis if all cranial sutures are clearly open. It is essential, however, that it is performed and evaluated by experienced clinicians. A three-dimensional CT scan images a craniosynostosis in a reliable way and is the most reliable objective diagnostic method.

A CT of the skull is always performed on suspicion of craniosynostosis. A three-dimensional CT scan will reliably diagnose craniosynostosis. In children with skull deformities moderate risk of craniosynostosis, an X-skull is indicated first, followed by a three-dimensional CT scan if the X-skull raises suspicion of craniosynostosis. On clear clinical suspicion of craniosynostosis high-risk craniosynostosis, an immediate three-dimensional CT scan is indicated, without an X-skull. Three-dimensional CT scanning should be reserved for suspected patients or for surgery planning.

Diagnostic imaging in the primary and secondary care sectors is advised against, unless performing skull x-rays has been approved by the craniofacial center and provided this does not lead to delay in referral. At suspicion of craniosynostosis this will be done at the shortest possible term. X-ray of the skull a-p, lateral, Towne of Tschebul is considered the first radiologic diagnostic test for craniosynostosis.

Ultrasound of the cranial sutures can be performed as an alternative to X-skull. Etiologic diagnostics and genetic counseling is the task of the clinical geneticist.

An etiologic or classifying diagnosis allows for making a prognosis not so much regarding the craniosynostosis, but rather regarding the child's general and psychomotor development and possible associated anomalies.
An etiologic diagnosis also provides for determining the risk of recurrence as well as the alternative choices in new pregnancies, such as prenatal diagnostics. Craniosynostosis is a birth defect that can present both in isolated form and syndromic form. In line with the international literature, we distinguish between nonsyndromic and syndromic craniosynostosis.

Dysmorphologic examination may distinguish between nonsyndromic and syndromic patients on the basis of the presence of dysmorphic characteristics. In addition, several bodily measurements are made and compared with reference values.

On indication, dysmorphologic examination of the whole patient is repeated. Possible indications are the development of new physical problems and newly observed delay in development in the course of time.

The January issue of the American Journal of Medical Genetics Part A is fully devoted to dysmorphologic examination of the face and extremities. The adequate performance and interpreting of dysmorphologic examination is one of the specific medical competencies of a clinical geneticist. Data of patients referred from through and of patients referred from through were compared on relative incidence of occurrence of the various types of craniosynostoses.

The total number of patients increased 1. The ratio nonsyndromic versus syndromic remained almost similar. In approximately 2 of every 15 children the craniosynostosis is of the syndromic type, in 1 in every 8 of them other than Apert, Crouzon or Saethre-Chotzen syndrome. The article does not make clear how syndromic and nonsyndromic craniosynostosis were distinguished.

The associated anomalies in the syndromic types of craniosynostosis notably were anomalies of the face and of the extremities. The London Dysmorphology Database version 1. More than half of the syndromes follow a Mendelian inheritance pattern, some are the result of teratogens including vitamin A and valproate.

Chromosomal disorders are seen in a proportion of patients with a craniosynostosis including 9p-, 11q-. Frias and Carey reviewed 4 population studies that established relations between the occurrence of minor anomalies and the risk of a major anomaly.

In the different studies, having 3 or more minor anomalies was associated with a From the same center in Paris, Lajeunie published a series of patients with a nonsyndromic scaphocephaly of a total of craniosynostosis-patients admitted between and Also the remaining literature does not provide information on the ratio between nonsyndromic versus syndromic scaphocephaly.

Lajeunie 33 reports that most patients of scaphocephaly and trigonocephaly are nonsyndromic. A striking finding was 4. In , Kan 34 described that in none of the 13 patients with scaphocephaly an FGFR2 mutation was found in a comprehensive screening of the whole gene. Butzelaar 35 describes a pilot study in 30 consecutive patients with scaphocephaly, which retrospectively analyzed how many patients had consulted the Clinical Genetics department, what genetic tests were used, and what the test results were.

In addition, the parents were sent a questionnaire on risk factors. Maternal alcohol use and smoking habit did not differ from those in the general population. Prematurity was more often seen in the study group than in the general population.

None of the 30 included patients had craniofacial dysmorphias. In 4 of the 30 patients another anomaly was diagnosed nevoid basal cell carcinoma syndrome, defects of the cardiovascular system and of the urinary tract. Zeiger 36 investigated genetic and environmental factors associated with a higher risk of scaphocephaly in a group of 42 children from craniofacial clinics in the Baltimore-Washington area.

Pathogenic mutations were not found. The same mutation occurred in the patient's nonafflicted father. The significance of this mutation is not clear. Of a series of patients admitted between and in Paris, Lajeunie published an analysis of patients with a trigonocephaly. Fifty-three Thirteen of these 53 had a known syndrome; the diagnosis in the other 40 patients was unknown.

Also in the group of trigonocephaly patients theman:woman ratio was skewed, that is, 3. The proportion of patients with a positive family history was 5. The proportion of twins in this series was 3 times higher than in the general population, and was higher than in the group of patients with a scaphocephaly 6. Azimi 40 investigated 25 patients with trigonocephaly diagnosed between and In 16 patients it was an isolated anomaly, 2 patients in addition had a craniosynostosis of the sagittal suture without associated anomalies.

Regarding the other 5 patients, in 1 the diagnosis of Say-Meyer trigonocephaly was made, in 1 I-cell disease and in another one Opitz-C syndrome.

A diagnosis could not be made in 2 patients. In 1 of 9 patients with a seemingly nonsyndromic trigonocephaly a mutation in the FGFR2 gene was found. Kress 42 found a FGFR1 mutation in 1 of 10 nonsyndromic trigonocephaly patients, who had or did not develop other dysmorphias. In , Kan 34 reported that in none of 17 patients with trigonocephaly a FGFR2 mutation was found in a comprehensive screening of the whole gene.

The patients were classified into groups, a group of 40 patients with an isolated trigonocephaly and a group of 36 patients with associated abnormalities. In 7 of the 36 syndromic trigonocephaly patients a 9p- or 11q-deletion was found.

Four of these deletions were not detectable with conventional cytogenetic analysis. In a case-report, Van der Meulen 44 describes a trigonocephaly in a patient with Muenke syndrome. In these types of craniosynostosis the coronal suture is involved; unilaterally in plagiocephaly, bilaterally in brachycephaly and oxycephaly. In oxycephaly there is also a craniosynostosis of the sagittal suture.

Mulliken 45 performed molecular diagnostics in patients with a bilateral coronal suture synostosis, of whom 38 had been diagnosed with Apert, Crouzon, or Pfeiffer syndrome and 19 had no specific diagnosis. In , Mulliken 46 reported results of molecular genetic diagnostics in 47 patients with a unilateral coronal suture synostosis. This was a prospective study in children admitted between and Thermanrwoman ratio was One patient
was clinically diagnosed with craniofrontonasal dysplasia.

Also the parents of the children were examined. Of the 47 children, 13 showed hypertelorism or had a parent with characteristics of craniosynostosis.

All above anomalies were found in these 13 children. The known craniosynostosis syndromes, Apert syndrome, Crouzon syndrome, Pfeiffer syndrome, Muenke syndrome, Saethre-Chotzen syndrome, Carpenter syndrome, and craniofrontonasal dysplasia CFND, are characterized by craniosynostosis of one or both coronal sutures next to other birth defects.

In addition, mutations were found in 7 new exons. In patients with a syndromic craniosynostosis, genetic diagnostics is performed on the guidance of the syndrome diagnosis.

FGFR1 mutations were not identified. Morriss-Kay 53 reported a prospective study in patients born between and Wilkie 52 tested patients in a diagnostic setting as from Submitting material of patients with a nonsyndromic scaphocephaly or trigonocephaly was strongly discouraged. If conventional molecular genetic diagnostics does not reveal mutations in one of the known craniosynostosis genes, additional diagnostics is warranted in patient of syndromic craniosynostosis.

Genetics diagnostics was performed with different techniques in the screening of 45 patients with a syndromic craniosynostosis without known mutation. The Crouzon and Pfeiffer syndromes cannot be easily distinguished clinically and genetically, but such distinction is not relevant to choice of treatment. The first choice for the genetic diagnostics is analysis of the FGFR2 gene.

Patients should be invited to contact the clinical geneticist again after reaching the age of 18 years to discuss the wish to have children and receive counseling if desired.

After referral of a child to a craniofacial team, the clinical geneticist of the team should be consulted. In addition, the family history regarding the occurrence of skull deformities and other birth defects should be documented. Molecular genetic diagnostics is not offered in patients with nonsyndromic scaphocephaly and nonsyndromic trigonocephaly. If these types of craniosynostosis occur in relatives, molecular genetic diagnostics is performed depending on the family history and preferably after consultation with a tertiary craniofacial center.

If a syndrome diagnosis has been made clinically, genetic diagnostics can be requested syndrome specifically. What organizational conditions should be minimally present for adequate and safe perioperative care for patients with a craniosynostosis?

Correction of craniosynostosis in childhood can cause relatively much blood loss. The risk of blood loss is higher in older patients and in corrective surgery of syndromic craniosynostosis. Next to the surgical and anesthesiologic challenges, we should take into account that syndromic patients may be associated with comorbidity. This is why optimal organizational conditions should be in place, before, during, and after the intervention.

This chapter deals with the specific risks involved in correction of nonsyndromic and syndromic craniosynostosis and recommendations are given to perform surgery as safely as possible.

All members of the multidisciplinary team should be aware of possible comorbidity, that is, mostly in case of a syndromic craniosynostosis with compromised airway with or without OSAS. It is recommended to set the threshold for postponing the intervention low in case of a recent upper airway infection in these patients described in Apert syndrome by Elwood 65 because airway infection is believed to be more strongly associated with complications.

A number of methods have been proposed to reduce both the blood loss and the need of homologous allogeneic blood transfusion in view of the risk of blood-transferable infection, immunologic reactions, coagulopathy, and transfusion-related acute lung injury TRALI , but most studies are not randomized double blind and have a B-C Level. These methods can be classified into different categories and are applied pre-, per- or postoperatively. In the polyclinic preoperative period:

Optimization of hematological conditions preoperatively, by administering erythropoietin EPO plus Fe supplement: A review concludes that multicenter studies are needed to determine the optimal dosing of EPO. Also the timing of administration, the optimal dosing of Fe, and cost-effectiveness must be determined.

Preoperative blood sampling for autologous transfusion during the operation: Opinions differ on this technique. Furthermore, the procedure is not child friendly as the newborns and infants must be anesthetized.

Preoperative surgical planning with the use of three-dimensional models seems to reduce operation time and thus also blood loss. Still, according to a C study, duration of the procedure is not decisive for the blood loss. Less invasive surgical procedures. Immediately before surgery the presence of blood products and availability of a pediatric intensive care unit PICU bed for the postoperative period should be confirmed in case significant blood loss related to the child's circulating capacity should occur.

A clear plan must have been made anticipating problems with intubation. The use of pre- and postoperative checklists and the time-out procedure TOP before start of surgery importantly add to safety of the entire perioperative process. Invasive monitoring is recommended in open procedures with expected severe blood loss. Invasive monitoring involves the use of a central venous line and an arterial line next to conventional monitoring capnography, ECG, pulse oxymeter, FiO2, temperature, and urine output and precordial Doppler.

The major goals of invasive monitoring are timely recognition of serious problems that may occur in open craniosynostosis surgery, such as hypohydration, hypotension, and electrolyte disturbances, as well as being able to check effectiveness of treatment of these problems.
Venous cerebral congestion by hyperflexion of rotation of the head must be avoided. Usually, the patient is placed in a moderate anti-Trendelenburg position to reduce blood loss cave air embolism, see below under Complications. Notably, in children with exorbitism the eyes should be well protected with eye cream, and pressure on the eyes must be avoided especially in prone position.

Single administration of an antibiotic, after the induction, before start of surgery is standard procedure. Most recommendations are aimed at a balanced technique that provides for cardiovascular stability with the use of opioids and volatile agents next to relaxants. The use of remifentanil infusion 0. An additional problem is that measurement of the lost volume intraoperatively is impeded by surgical technique and type of patient. Few studies have been performed to optimize blood loss measurement and findings are mostly not conclusive.

Infiltration of the skin with vasoconstrictors before the incision is much used but its effectiveness is disputed because of the greater degree of bleeding with treatment of the peristium and bone. Controlled hypotension is rarely used because it has no clear benefits and can be disadvantageous in case of ICP and also in the anti-Trendelenburg position.

The acute normovolemic hemodilution ANH technique is used before the surgical intervention after the child is brought under anesthesia. It involves removal of whole blood via the arterial or central line which is replaced with colloids or with crystalloids according to precise calculations and formulas. According to a B study, this technique in itself is not sufficient to reduce or avoid homologous blood transfusion in this type of surgery.

Duncan 82 found no difference in allogeneic transfusion rate when a cell saver was used and when not. Intraoperative antifibrinolytics. Aprotinin is no longer used after serious complications in cardiac surgery. Tranexamic acid: its use in itself is controversial, but it is has been applied in combination with other methods.

Guidelines for the application of blood products are in place in every hospital as established by the local transfusion committees, and in conformity with the CBO Guideline Blood transfusion Venous air embolism VAE and the subsequent cardiovascular collapse can be prevented by a precise technique and by rapid application of adequate monitoring precordial Doppler, capnography, echocardiography, transcutaneous O2—CO2 monitoring, esophageal stethoscope, and venous central line.

Although Faberowski 86 reports an incidence of Meyer 88 reports a 2. The hemodynamic consequences of VAE are generally insubstantial, provided the patients are well monitored and measures are taken upon signs of VAE. As a result a massive blood loss the patient may develop relevant consumption coagulopathy and dilutional coagulopathy characterized, in principle, by depletion of soluble clotting factors.

The situation changes when these children undergo a distraction osteotomy procedure and need to be intubated acutely in case of respiratory insufficiency, or when the distraction materials are being removed.

Evidence from the C level studies is conflicting on this issue. One of the 2 studies recommends immediate fiberoptic intubation, whereas the other study claims that the distractors exert minimal effect in the anesthesiologic conditions when certain factors must be taken into account the right screwdrivers and cutting pliers must always be available and that removal of the vertical bar allows for direct laryngoscopy.

The possibility of cerebral salt wasting syndrome should be considered when a patient develops hypotension after craniosynostosis surgery. Criteria for extubation at the end of the procedure, before transport to the ICU, are the following: rapid recovery of spontaneous and stable breathing, hemodynamic stability, normothermia, short to medium operation time with relatively little blood loss, and no continuous large blood loss through the surgical drains.

In all studies recommend ICU admission after open craniosynostosis surgery to continue volume management there and, if necessary, start artificial respiration. Most studies on postoperative transfusion management report a tendency to overtransfusion. There is scarce literature on postoperative pain treatment after craniosynostosis surgery.

In Rotterdam 2 double blind, placebo-controlled studies have investigated pain treatment in the first 24 hours after cranial correction for craniosynostosis. Van der Marel 95 treated 20 children with oral paracetamol and 20 with rectal paracetamol. The pain scores in the oral group were higher, but this effect disappeared with exclusion of the patients who had vomited after oral administration.

Despite the fact that Rectal administration of paracetamol was recommended. Prins 96 describes 12 children who received intravenous paracetamol and 14 children who received rectal paracetamol. This is indicative of more discomfort, possibly caused by more pain, and intravenous administration was recommended. In the European literature, the use of paracetamol and NSAIDs is recommended if the blood loss is not significant with or without codeine phosphate British literature.

All textbooks emphasize, and this also appears from our own practice, is that the pain scores are surprisingly low. In patients operated on for nonsyndromic craniosynostosis, invasive monitoring can usually be discontinued the following day.

Despite the proven effectiveness of erythropoietin and blood drawing for autologous transfusion, the use of these strategies is discouraged as they involve high costs and require repeated venipuncture, which is not child friendly.

Introduction of less invasive interventions is associated with less blood loss. As these, however, are very young children with a smaller circulating capacity, blood loss relatively is still significant. In selected cases admission to a medium care unit could be considered defined as a monitored bed providing for artificial respiration. The anesthesiologic goals in craniofacial surgery can only be realized in specialized pediatric centers, where multidisciplinary perioperative care is provided by a team composed of plastic surgeon, neurosurgeon, maxillofacial surgeon, pediatrician, pediatric anesthesiologist, pediatric intensivist, and specialized pediatric nurses with experience and means to manage and monitor this type of patients and where a sufficient number of children is operated on to keep the experience of the team at a high level.
Administration of EPO preceding the intervention, as well as collecting autologous blood for autotransfusion are advised against. Postoperatively a bed in a PICU must be available.

In less drastic interventions medium care may perhaps suffice, although 1 should be aware that these are mostly very young children with a smaller circulating capacity, in whom even slight postoperative blood loss must be monitored carefully.

Invasive monitoring is recommended in the case of open procedures with expected severe bleeding. Overtransfusion in the postoperative phase should be prevented by adhering to the guideline on transfusion management. Extubation at the end of the operation, before transport to the ICU, is possible in case of rapid recovery of spontaneous and stable breathing, hemodynamic stability, normothermia, short to medium operation time with relatively little blood loss, and no continuous large blood loss through the surgical drains.

The analgesic of choice for postoperative pain treatment is paracetamol. If this is not sufficient, an NSAID can be added even though this could increase the risk of postoperative bleeding.

What are the indications for surgery in the different types of nonsyndromic craniosynostosis? What treatment is most indicated for the different types of nonsyndromic craniosynostosis?

Nonsyndromic craniosynostosis can present in varying severity of the deformity of the skull. Surgical treatment seems indicated on the basis of:

The variability in both the severity of the morphologic abnormality and in the occurrence of increased ICP may occasionally result in less strict operative indication on the basis of these parameters. Many different surgical techniques have been described for the treatment of nonsyndromic craniosynostosis but the functional results of these techniques have not always been shown convincingly.

In addition, the timing of surgery is debated. For patients with a synostosis of multiple cranial sutures in whom a syndromic diagnosis cannot yet be made, the method of treatment of syndromic craniosynostosis is recommended.

This chapter will place a focus on craniosynostosis of the sagittal suture scaphocephaly, metopic suture trigonocephaly, unilateral coronal suture frontal plagiocephaly, and lambdoidal suture pachycephaly. The indications for treatment of craniosynostosis are the risk of ICP and the morphologic abnormality of the skull and face. The chapter on cognition and behavior provides evidence of absence of a relation between cognitive functions in children with a nonsyndromic craniosynostosis and surgical treatment or not, severity of the deformity, or age at operation.

The prevention or treatment of the associated neuropathology is therefore no indication for surgery. The morphology can vary from very mild to an evidently abnormal skull shape. In sagittal suture synostosis, compensatory growth of the other cranial sutures results in increasing deformity in the course of time, so that, for example, the frontal bossing in scaphocephaly can intensify after birth.

Barritt evaluated a series of 44 children with scaphocephaly, of whom 34 did not undergo surgery. This was common policy in this clinic at the time for children presenting with this condition at an age older than 6 months. In these 34 children, the deformity in any patient did not seem to improve in the first 10 years. The presentation of trigonocephaly also varies from very mild to severe.

Its mildest form is characterized only by a bony ridge as a result of premature fusion of the metopic suture. In follow-up, cosmetic worsening is only rarely seen. The other end of the spectrum is characterized by a wedge-shaped forehead with hypotelorism and supraorbital retrusion on the basis of decreased growth of the forehead widthwise. Frontal plagiocephaly also shows some variation in phenotype, notably in facial asymmetry. Unilateral synostosis of a lambdoidal suture causes increasing asymmetry of the back of the head and particularly also of the face.

Increased ICP does not seem to be prominent in this type of synostosis, although reliable research on this issue is lacking see chapter 8. Since the first operative intervention for craniosynostosis, many surgical techniques for the various types of craniosynostosis have been described.

A broad distinction is made between osteoclastic techniques and remodeling techniques. In the first group bone is removed, enabling the developing and expanding brain to change the shape of the skull, in part because the adverse impulse to the growth direction of the skull also is removed. From this technique evolved the remodeling techniques, because it was suspected that one cannot only rely on the self-correcting capability of the skull and the brain.

These techniques are aimed at directly achieving the desired skull shape by a kind of reconstruction. Randomized comparisons of the different surgical techniques have never been performed.

For the different types of nonsyndromic craniosynostosis we will provide a review of the literature regarding the different techniques in these 2 groups, but also pay attention to the new developments in the past 10 years in a separate section. Ingraham described a technique using bilateral parasagittal strip craniectomies, in which the fused suture was not resected.

To prevent the problem of premature reossification of the skull bones he applied a layer of plastic to the bone rims. Successful treatment required early intervention, however, preferably in the first 2 months of life. The results of this technique were confirmed by other authors. Albright presented a sagittal strip craniectomy with biparietal wedge osteotomies. The same procedure was performed by Marucci in 89 patients, but they noticed that 7 of the 89 patients later developed a cosmetically unsatisfactory vertex bulge, which they interpreted as a sign of possibly raised ICP or a new synostosis.

Also Florisson reported that some patients with scaphocephaly again showed signs of raised ICP postoperatively. Amm presented a technique, in which the usual 2. They explicitly try to make use of gravitational force by keeping the infant strictly in supine position during sleep for a period of 3 weeks. Using this technique an average improvement of the CI of 8. All in all, many of the described techniques represent only slight modifications of previously presented techniques and not real conceptual changes.
The sagittal suture itself is not resected, and the temporoparietal bone is bent outwards. In this technique, the anteroposterior diameter of the skull is dynamically shortened by attaching the bone strip in the middle of the skull, which is shortened, to the frontal bone. Thus, it is in fact also a compression technique, which might be associated with an increase in ICP. Only a global development quotient, however, was tested in very young patients, without inclusion of a control group, which raises great doubt on the reliability of this finding.

In older children or in children with a more pronounced dysmorphology notably frontal bossing, a more extensive calvarial reconstruction is needed, with resection and reconstruction of the frontobiparietal complex.

The authors concluded that in this population this more extensive procedure had no adverse effect on postoperative growth rate of the skull or on intracranial volume. Lauritzen reported in on the first operations in which distraction was used to correct craniosynostosis.

This method is evidently suitable for scaphocephaly in view of the unidirectional growth delay widthwise. The results are not worse, but also not better than those of the current techniques and the small numbers preclude valid comparison.

Also regarding the newer surgical techniques, randomized or comparative studies have not been published. Also in view of the complex three-dimensional dysmorphology seen in metopic synostosis, treatment by sutuectomy solely is considered insufficient. Posnick analyzed the growth process on the basis of CT data in 10 patients and concluded that correction of the hypotelorism had remained insufficient. Selber found that the use of interpositional bone grafts in a series of 68 metopic children led to a decrease in the development of temporal dents.

In a population of 92 trigonocephaly children in which no bone graft was used, Van der Meulen, however, observed an automatic growth correction of the hypotelorism on x-ray, on account of which the use of an interpositional bone graft was judged to be unnecessary. From a photographic evaluation of 45 patients, Hilling concluded that a satisfactory esthetic outcome on the long term was largely dependent on a good initial reconstruction.

A distinction is made between unilateral and bilateral frontal advancement techniques. Some surgeons prefer the unilateral advancement technique so as to be able use the unaffected side as a reference for the degree of advancement, whereas others maintain that adequate advancement with good symmetry is only possible if the entire frontal bone and both orbital margins have been dissected.

Barone and Jimenez report since the late s on endoscopic strip craniectomy of the affected suture. The benefits mainly lie in low morbidity, short hospitalization, and little need of blood transfusions in comparison with the classic fronto-orbital advancement technique. The cosmetic outcome is reported as good, but it not adequately quantified. For this reason, the technique was adapted to an endoscopic unilateral fronto-orbital advancement technique.

This was applied in 10 patients, who did not develop any complications, did not need blood transfusions, and could be discharged home after 2 days. The cosmetic outcome was assessed to be similar to that of fronto-orbital advancement. The number of publications on lambdoid suture synostosis is very limited, probably because of the very low prevalence.

Unilateral lambdoid suture synostosis causes only relatively little volumetric restriction. The morphologic changes consist of an asymmetry of the face and an asymmetric basal occipital region. Kim reports a study comparing distraction versus remodeling surgery.

Among the patients were 3 children with brachycephaly treated with the distraction method and 4 with remodeling. The distraction method led to satisfactory calvarial expansion and an esthetically pleasing outcome. Advantages of distraction are the significantly shorter operation time, less bleeding, shorter stay at the neonatal intensive care unit NICU, and better skull shape. A disadvantage of the distraction method is that children must have reached the age of 6 months.

The authors are of the opinion that these downsides do not outweigh the benefits of distraction. Most children with a craniosynostosis are operated on in the first year of life. A direct evaluation of surgical outcomes is possible, but a final evaluation is not possible until the child has fully grown at adult age.

As a surgeon's career spans perhaps 25 year, and at most 30 years, the surgeon can only at an advance career stage assess the result of his or her own work. There may be a tendency to ascribe a poor result to a wrongly performed surgical technique, without questioning the technique itself. Furthermore, knowledge about the natural course of the condition with and without surgical intervention is usually lacking. This shortcoming undoubtedly is coresponsilble for the fact that we see regular shifts in type of intervention in the course of decades and that paradigms change.

One and the same research group may recommend a certain approach in the 1 decade and disassociate oneself from it in the next decade. Only a few solid objective results with a long follow-up are available, whereas comparative studies of different surgical techniques with or without randomization have not at all been performed. Moreover, notably the larger series yield mixed data, because all craniosynostoses, including the syndromic types, are pooled. One way to assess the outcome of surgery is looking at the operative morbidity and mortality.

Most series are too small, however, to yield significant data. Morbidity is reflected in the complications that may occur and length of hospitalization. Although the first is a relatively objective score, the second is certainly not. Length of hospitalization strongly depends on the treating physicians, but also on the healthcare system itself. To illustrate this, for similar procedures the average length of hospitalization in North America is significantly shorter than in Europe.

The patients are simply discharged home earlier, which possibly is enabled by a better outpatient care system. Generally, hospitalization after minimally invasive interventions is 1 or 2 days; for more complex interventions 2 to 7 days. As postoperative infections are very rare, only large series can yield significant data.
In series of patients who underwent surgery for unisutural craniosynostosis only 2.0. In general, the complications rates in the nonsyndromic craniosynostoses are significantly lower than in the syndromic craniosynostoses, 3.

The use of metal plates and screws has been customary in a certain period. Meanwhile, it has become evident that application of this material in young children may lead to intracranial translocation of the plates and screws, with the screw ends penetrating the dura.

In the past few years, resorbable plates and screw devices have been applied for specific indications in craniosynostosis surgery.

The outcome of surgery can also be expressed by need for secondary revision surgery. This is rarely needed. Amm routinely uses three-dimensional surface analysis software to analyze CT-scans.

Computerized tomography scanning, however, is a source of radiation exposure. Still it enables to assess the outcome in a relatively reliable and objective manner, and this yielded a mean 8. Marucci noticed a cosmetically unsatisfactory vertex bulge after minor surgery in 7 of 89 patients. At follow-up in the course of years after reconstruction of trigonocephaly, the lateral sides of the forehead often appear to revert to a certain degree and temporal depressions may be seen, which however seldom are cosmetically unsatisfactory to the degree that revision surgery is justified.

Hilling evaluated the result of bifrontal-orbital advancement in trigonocephaly from photographs of the patients pre- and postoperatively and after follow-up by a panel of independent assessors with the aid of a scoring form.

They concluded that the cosmetic result generally was good to very good, but also that the severity of the deformity does not influence the eventual outcome. In addition, they established that the obtained result is stable during time and independent of the age at operation 6 to 15 months. Temporal depressions are ultimately the most frequent abnormalities at follow-up and cosmetically usually only little unfavorable. Van der Meulen performed standard x-rays of the skull before and after operation and in the course of time and noted that although in fronto-orbital advancement, the hypotelorism had not been actively corrected, autocorrection still occurred during time.

Assessment of the result of the various techniques applied in frontal plagiocephaly is highly subjective. Objective outcome measures are lacking and assessment therefore mainly consists of an evaluation of the achieved facial symmetry. Two of those patients underwent corrective ophthalmological surgery for strabismus as a result of a malformed orbit, whereas a third patient had rejected the same proposed intervention.

The earlier mentioned panel assessment of photographs by Hilling in trigonocephaly, was also applied by the same researchers in 59 patients with frontal plagiocephaly. Also in this deformity, the temporal depression is the most striking finding at follow-up. Direct evidence for this supposition, however, is lacking. For that matter, this research group also concludes that the frequency of temporal depressions decreases with increasing experience of the surgeon.

Lo specifically assessed the orbital morphology both of bone and soft tissue after surgical intervention and evaluated this by means of CT-scans. They noted that the orbital morphology had genuinely improved, but also that complete symmetry had not been obtained.

Furthermore, outgrowth, however, was normalized. The timing of surgery differs worldwide and is dependent, among other things, on the surgical technique used. In patients with boat-shaped head, it was observed that the initially improved CI index had slightly decreased 1 year after correction, whereas in fronto-orbital advancements, the frontolateral skull will again slightly receed a part reversal to the primary deformity. This in general does not cause cosmetic problems, the more so because some surgeons in anticipation will apply overcorrection.

Thompson proposes that nonsyndromic craniosynostosis ideally is corrected within the first year of life, with scaphocephaly as a possible exception, corrected with early modified strip craniectomy. The largest series is reported by Marchac, nearly patients with all types of craniosynostoses were operated on in Paris.

It is suggested to correct brachycephaly at the age of 2 to 4 months often syndromic patients with raised ICP and the other conditions at the age of 6 to 12 months. The reason why is not argued, however. Amm prefers correction of scaphocephaly at the age of 6 to 12 months, without clear argumentation. Fearon, evaluating 16 children with trigonocephaly, established that the younger the child, the more spontaneous improvement of the hypotelorism after surgery, even though the hypotelorism itself was not surgically corrected.

In , they concluded that surgery at the age of 3 months would be ideal for good development of the orbits. Thirteen years later, however, the same research group concludes that the younger the child, the larger the growth inhibition of the skull after operative intervention.

Their timing then is 4 months for the scaphocephalic child to prevent progression of the frontal bossing and 9 months for monosutural craniosynostoses requiring advancement so that in any case the frontal band is strong enough, but the dural capacity of bone regeneration also is still sufficient to close all holes.

Also for frontal plagiocephaly, there is no consensus on the timing of surgery. Although minimal invasive surgical techniques need to be performed at young age, ages of 6 months to from 15 to 18 months are recommended for major reconstruction. Here, too, the most convincing argument for late surgery at the age of 18 months is a possibly lower chance of later relapse. Although the risk of relapse seems to be somewhat increased after an early limited intervention, the risk is probably small.

This should be weighed against the burden of a late complete cranial remodeling with longer operation time and more bleeding. This guideline therefore does not pronounce on a preference for early or late surgery. Still, in view of the results of Renier regarding the outcome of surgery after the first year of life, it is recommended to perform the intervention in the first year of life.

Sub group — a included 11 patients within age group 18—24 months, and sub—group — b included 09 patients within age group 25—36
months. Informed consent was obtained from all participating adult subjects, and parents, guardians of minors or incapacitated adults. None of the children were known to be mentally retarded or have associated syndromes.

Exclusion criteria were submucous cleft palate, identified syndrome, and hearing loss sensorial or persistent conductive hearing loss despite tympanostomy. Palatostal fistulas were recorded—both anterior and posterior fistulas. All pre- and post-operative examinations were carried out by the same surgeon, otolaryngologist, and speech therapist. A total of 20 patients underwent Pinto’s modification of Wardill—Kilner palatoplasty without radical dissection of the levator veli palatini LVP and tensor veli palatini TVP muscle.

Twenty patients in the age group of 18—36 months underwent Pinto’s modification of Wardill—Kilner two-layer palatoplasty without radical dissection soft palate musculature.

The Hamular process was fractured in all these patients to free the tensor veli palatine tendon to facilitate the posteromedial displacement of velar muscles and the mucoperiosteal flaps. Anchoring sutures-nasal layer was closed using 3—0 catgut sutures. The same sutures were passed through the oral layer later as anchoring sutures. This minimized the dead space and prevented the falling of the flap. At the end 1 or 2 stay sutures were placed for the lateral releasing incision.

All the patients were given postoperative Amox-clav antibiotics for 7 days and discharged after a week. All the children were regularly seen at 1-month interval for 1 year by the surgeons and the speech therapist. All the children underwent speech assessment and the counseling. In all the patients following variable were determined: quality of speech, incidence of fistula, and age at the time of surgery.

Clinically significant fistulas were determined by the presence of either hypernasal speech, articulation problem or fluid regurgitation from the nose. Speech was classified into three different groups, namely A nasality, B articulation, C intelligibility, each of which is further subdivided by numbers.

Nasality was divided into A1 normal, A2 mild hyper nasality, A3 moderate hyper nasality, A4 severe hyper nasality. Articulation was divided into B1 normal, B2 one to two consistent errors only with no deterioration in speech, B3 one to two consistent error with deterioration in connected speech or three or more errors but intelligible, B4 multiple errors, frequently unintelligible.

Intelligibility was graded into- C1 intelligible at all times, C2 sometimes unintelligible, C3 unintelligible most of the time. Velopharyngeal incompetence was diagnosed clinically by the surgeons and the speech therapist.

Twenty patients were enrolled in the study, and all attended regular follow-up till 6 months postoperatively. There were 12 males and 8 females. The age at repair ranged from 18 months to 36 months mean age No major perioperative or postoperative complications occurred throughout the study except in one patient who had secondary bleeding after 48 hours of the surgery.

Many patients showed improvement in their 6-month postoperative speech in both groups. The preoperative values of speech assessment all three parameters, the value indicated that two-layer palatoplasty modified Wardill—Kilner V-Y pushback technique without an intravelar veloplasty technique was good for speech.

Postoperative palatal fistulas were encountered in four patients in the study. In , Johann Friedrich Dieffenbach, the director of the Clinical Institute of Surgery at Charite Hospital in Berlin, first described closure of the hard palate by mucosal elevation.

In , he advanced palatal surgery even further with the introduction of relaxing incisions to ease palatal closure. After his death in , Dieffenbach was succeeded as the director of the Clinical Institute of Surgery by Bernhard von Langenbeck, who also became a leading innovator in cleft repair.

In the early s, soft palate closure was also being studied. Leonard Furlow later advanced cleft palate repair surgery with the introduction. The major criteria for determining the success of cleft palate repair are subsequent speech development, maxillofacial growth, and complete closure of palatal defect.

There is no question that speech development is the major goal of palatoplasty and therefore success or failure of this operation is usually measured by the proportion of patients with normal speech versus patients with remaining hypernasality. The question which persists is the timing of palatoplasty and its correlation with normal speech production.

There is evidence that the earlier the palatal repair is performed the better the speech result one can expect. However there are objections to palatoplasty because of the previously mentioned concept that cleft palate repair is the major cause of mid-facial growth inhibition and secondary maxillofacial deformities. Bardach's and Salyer's long-term clinical observation indicate that palatoplasty cannot be considered the only cause of mid-facial growth aberrations and inhibition.

Cleft palate surgery is one of the greatest challenges for oral and maxillofacial surgeons. A good result requires an esthetic functional closure without impairment of facial growth, allowing normal speech development. Speech is a complex phenomenon that is best learned once and the younger the better. There are still no standard protocols to address the issues of ideal timing for cleft palate repair to attain optimal speech and to avoid abnormal maxillofacial growth after repair. While there are many controversies on the timing of cleft palate surgery, the current debate concerns how early palatal repair should be performed.

The ideal timing of cleft palate closure should depend upon the type of cleft involved, the patient's condition and the capabilities of the cleft team to manage associated morbidities. Because some cleft patients have associated anomalies and syndromes, the timing of palatoplasty should be tailored individually after thorough clinical evaluation. They concluded that there was highly significant improvement in hypernasality, nasal emission, and velopharyngeal closure. Severity of the SMCP did not correlate with the degree of preoperative speech abnormality but was a significant predictor of outcome of surgery, with less severe total SMCP score of 0—3 having less satisfactory end results and lesser degrees of
In our study the Wardill—Kilner technique gives satisfactory results in terms of velopharyngeal function, so velar surgery was always worthwhile for cleft palate repair. Improvement was nondependent variable with regard to cleft types. In the study continued by Grobbelaar et al. Similarly in our study the speech results were significantly much better in the younger age group 18—24 months than our older age group 24—36 months.

Heliovaara et al. They compared incidence of palatal fistula in their patients who had undergone palatoplasty. In our study we found an 8.

Postpalatoplasty speech results can be affected by the length of the palate, the active mobility of the palatal muscles, the depth of the nasopharynx, the presence of adenoidal tissues, and the occurrence of palatal fistulae or gapping. The art of cleft palate repair has enjoyed considerable development over many years. Although the controversies regarding the timing and closure of a cleft palate seem to have been resolved, with a consensus for surgery being completed at 18 months, there are still many issues which need to be resolved by well-controlled, randomized, prospective clinical trials to ascertain the optimal timing of palatoplasty, and its long-term relationships with speech development and maxillofacial growth.

Results from our study indicate that two-layer palatoplasty modified Wardill—Kilner V-Y pushback technique without an intravelar veloplasty technique was good for speech. Source of Support: Nil. Conflict of Interest: None declared. National Center for Biotechnology Information, U.


About Cleft-Palate And Hare-Lip: The Earlier Operation On The Palate (Classic Reprint) Writer

In children with skull deformities moderate risk of craniosynostosis, an X-skull is indicated first, followed by a three-dimensional CT scan if the X-skull raises suspicion of craniosynostosis. On clear clinical suspicion of craniosynostosis high-risk craniosynostosis, an immediate three-dimensional CT scan is indicated, without an X-skull.

Three-dimensional CT scanning should be reserved for suspected patients or for surgery planning. It appeared that ultrasound could well visualize a craniosynostosis in 26 children with craniosynostosis aged from 2 to 7 months and in 23 of the 24 children with craniosynostosis aged from 1 to 11 months. The clinical diagnosis of craniosynostosis by means of physical examination forms the basis.

Even though an X-skull is not always reliable, notably at very young age several months old, as first further diagnostic test, it provides much information and can exclude craniosynostosis if all cranial sutures are clearly open.

It is essential, however, that it is performed and evaluated by experienced clinicians. A three-dimensional CT scan images a craniosynostosis in a reliable way and is the most reliable objective diagnostic method. A standardly performed three-dimensional CT scan to objectivize the deformity is highly recommended for operative planning. X-skull a-p, lateral, Towne for back of the head, Tschebul for forehead is always performed in suspected craniosynostosis.

If unclear, because of very young age of the patient, it is recommended to repeat X-skull after 1 or 2 months. If X-skull confirms, or does not exclude, a synostosis, a CT-scan with three-dimensional-reconstruction is performed. In case of very strong suspicion of craniosynostosis, the X-skull is skipped and a CT-scan with three-dimensional reconstruction is performed without delay.

Ultrasound of the cranial sutures can be performed as an alternative to X-skull. Etiologic diagnostics and genetic counseling is the task of the clinical geneticist. An etiologic or classifying diagnosis allows for making a prognosis not so much regarding the craniosynostosis, but rather regarding the child's general and psychomotor development and possible associated anomalies.

An etiologic diagnosis also provides for determining the risk of recurrence as well as the alternative choices in new pregnancies, such as prenatal diagnostics. Craniosynostosis is a birth defect that can present both in isolated form and syndromic form. In line with the international literature, we distinguish between nonsyndromic and syndromic craniosynostosis.

Dysmorphologic examination may distinguish between nonsyndromic and syndromic patients on the basis of the presence of dysmorphic characteristics.

In addition, several bodily measurements are made and compared with reference values. On indication, dysmorphologic examination of the whole patient is repeated. Possible indications are the development of new physical problems and newly observed delay in development in the course of time. The January issue of the American Journal of Medical Genetics Part A is fully devoted to dysmorphologic examination of the face and extremities.

The adequate performance and interpreting of dysmorphologic examination is one of the specific medical competencies of a clinical geneticist. Data of patients referred from through and of patients referred from through were compared on relative incidence of occurrence of the various types of craniosynostoses.

The total number of patients increased 1. The ratio nonsyndromic versus syndromic remained almost similar. In approximately 2 of every 15 children the craniosynostosis is of the syndromic type, in 1 in every 8 of them other than Apert, Crouzon of Saethre-Chotzen syndrome.

The article does not make clear how syndromic and nonsyndromic craniosynostosis were distinguished. The associated anomalies in the syndromic types of craniosynostosis notably were anomalies of the face and of the extremities.
The London Dysmorphology Database version 1. More than half of the syndromes follow a Mendelian inheritance pattern; some are the result of teratogens including vitamin A and valproate. Chromosomal disorders are seen in a proportion of patients with a craniosynostosis including 9p-, 11q-. Frias and Carey reviewed 4 population studies that established relations between the occurrence of minor anomalies and the risk of a major anomaly.

In the different studies, having 3 or more minor anomalies was associated with a From the same center in Paris, Lajeunie in published a series of patients with a nonsyndromic scaphocephaly of a total of craniosynostosis-patients admitted between and Also the remaining literature does not provide information on the ratio between nonsyndromic versus syndromic scaphocephaly.

Lajeunie 33 reports that most patients of scaphocephaly and trigonocephaly are nonsyndromic. A striking finding was 4. In , Kan 34 described that in none of the 13 patients with scaphocephaly an FGFR2 mutation was found in a comprehensive screening of the whole gene.

Butzelaar 35 describes a pilot study in 30 consecutive patients with scaphocephaly, which retrospectively analyzed how many patients had consulted the Clinical Genetics department, what genetic tests were used, and what the test results were. In addition, the parents were sent a questionnaire on risk factors. Maternal alcohol use and smoking habit did not differ from those in the general population.

Prematurity was more often seen in the study group than in the general population. None of the 30 included patients had craniofacial dysmorphias. In 4 of the 30 patients another anomaly was diagnosed nevoid basal cell carcinoma syndrome, defects of the cardiovascular system and of the urinary tract. Zeiger 36 investigated genetic and environmental factors associated with a higher risk of scaphocephaly in a group of 42 children from craniofacial clinics in the Baltimore-Washington area.

Pathogenic mutations were not found. The same mutation occurred in the patient's nonafflicted father. The significance of this mutation is not clear. Of a series of patients admitted between and in Paris, Lajeunie published an analysis of patients with a trigonocephaly.

Fifty-three-Three. Thirteen of these 53 had a known syndrome; the diagnosis in the other 40 patients was unknown. Also in the group of trigonocephaly patients thennuromann ratio was skewed, that is, 3. The proportion of patients with a positive family history was 5. The proportion of twins in this series was 3 times higher than in the general population, and was higher than in the group of patients with a scaphocephaly 6. Azini 40 investigated 25 patients with trigonocephaly diagnosed between and In 16 patients it was an isolated anomaly, 2 patients in addition had a craniosynostosis of the sagittal suture without associated anomalies.

Regarding the other 5 patients, in 1 the diagnosis of Say-Meyer trigonocephaly was made, in 1 I-cell disease and in another one Optiz-C syndrome. A diagnosis could not be made in 2 patients. In 1 of 9 patients with a seemingly nonsyndromic trigonocephaly a mutation in the FGFR2 gene was found. Kress 42 found a FGFR1 mutation in 1 of 10 nonsyndromic trigonocephaly patients, who had or did not develop other dysmorphias.

In , Kan 34 reported that in none of 17 patients with trigonocephaly a FGFR2 mutation was found in a comprehensive screening of the whole gene. The patients were classified into groups, a group of 40 patients with an isolated trigonocephaly and a group of 36 patients with associated abnormalities. In 7 of the 36 syndromic trigonocephaly patients a 9p- or 11q-deletion was found. Four of these deletions were not detectable with conventional cytogenetic analysis.

In a case-report, Van der Meulen 44 describes a trigonocephaly in a patient with Muenke syndrome. In these types of craniosynostosis the coronal suture is involved; unilaterally in plagiocephaly, bilaterally in brachycephaly and oxycephaly.

In oxycephaly there is also a craniosynostosis of the sagittal suture. Mulliken 45 performed molecular diagnostics in patients with a bilateral coronal suture synostosis, of whom 38 had been diagnosed with Apert, Crouzon, or Pfeiffer syndrome and 19 had no specific diagnosis. In , Mulliken 46 reported results of molecular genetic diagnostics in 47 patients with a unilateral coronal suture synostosis.

This was a prospective study in children admitted between and The meromann ratio was One patient was clinically diagnosed with craniofrontonasal dysplasia. Also the parents of the children were examined. Of the 47 children, 13 showed hypertelorism or had a parent with characteristics of craniosynostosis. All above anomalies were found in these 13 children. The known craniosynostosis syndromes, Apert syndrome, Crouzon syndrome, Pfeiffer syndrome, Muenke syndrome, Saethre-Chotzen syndrome, Carpenter syndrome, and craniofrontonasal dysplasia CFND , are characterized by craniosynostosis of one or both coronal sutures next to other birth defects.

In addition, mutations were found in 7 new exons. In patients with a syndromic craniosynostosis, genetic diagnostics is performed on the guidance of the syndrome diagnosis. FGFR1 mutations were not identified.

Morris-Kay 53 reported a prospective study in patients born between and Wilkie 52 tested patients in a diagnostic setting as from Submitting material of patients with a nonsyndromic scaphocephaly or trigonocephaly was strongly discouraged.

If conventional molecular genetic diagnostics does not reveal mutations in one of the known craniosynostosis genes, additional diagnostics is warranted in patient of syndromic craniosynostosis. Genetics diagnostics was performed with different techniques in the screening of 45 patients with a syndromic craniosynostosis without known mutation. The Crouzon and Pfeiffer syndromes cannot be easily distinguished clinically and genetically, but such distinction is not relevant to choice of treatment.

The first choice for the genetic diagnostics is analysis of the FGFR2 gene. Patients should be invited to contact the clinical geneticist again after reaching the age of 18 years to discuss the wish to have children and receive counseling if desired.

After referral of a child to a craniofacial team, the clinical geneticist of the team should be consulted. In addition, the family history regarding the
occurrence of skull deformities and other birth defects should be documented.

Molecular genetic diagnostics is not offered in patients with nonsyndromic scaphocephaly and nonsyndromic trigonocephaly. If these types of craniosynostosis occur in relatives, molecular genetic diagnostics is performed depending on the family history and preferably after consultation with a tertiary craniofacial center. If a syndrome diagnosis has been made clinically, genetic diagnostics can be requested syndrome specifically. What organizational conditions should be minimally present for adequate and safe perioperative care for patients with a craniosynostosis?

Correction of craniosynostosis in childhood can cause relatively much blood loss. The risk of blood loss is higher in older patients and in corrective surgery of syndromic craniosynostosis. Next to the surgical and anesthesiologic challenges, we should take into account that syndromic patients may be associated with comorbidity. This is why optimal organizational conditions should be in place, before, during, and after the intervention.

This chapter deals with the specific risks involved in correction of nonsyndromic and syndromic craniosynostosis and recommendations are given to perform surgery as safely as possible. All members of the multidisciplinary team should be aware of possible comorbidity, that is, mostly in case of a syndromic craniosynostosis with compromised airway with or without OSAS.

It is recommended to set the threshold for postponing the intervention low in case of a recent upper airway infection in these patients described in Apert syndrome by Elwood 65 because airway infection is believed to be more strongly associated with complications.

A number of methods have been proposed to reduce both the blood loss and the need of homologous allogeneic blood transfusion in view of the risk of blood-transferrable infection, immunologic reactions, coagulopathy, and transfusion-related acute lung injury TRALI, but most studies are not randomized double blind and have a B-C Level. These methods can be classified into different categories and are applied pre-, peri- or postoperatively.

In the polyclinic preoperative period: Optimalization of hematological conditions preoperatively, by administering erythropoietin EPO plus Fe supplement: A review concludes that multicenter studies are needed to determine the optimal dosing of EPO.

Also the timing of administration, the optimal dosing of Fe, and cost-effectiveness must be determined. Preoperative blood sampling for autologous transfusion during the operation: Opinions differ on this technique.

Furthermore, the procedure is not child friendly as the newborns and infants must be anesthetized. Preoperative surgical planning with the use of three-dimensional models seems to reduce operation time and thus also blood loss.

Still, according to a C study, duration of the procedure is not decisive for the blood loss. Less invasive surgical procedures. Immediately before surgery the presence of blood products and availability of a pediatric intensive care unit PICU bed for the postoperative period should be confirmed in case significant blood loss related to the child's circulating capacity should occur.

A clear plan must have been made anticipating problems with intubation. The use of pre- and postoperative checklists and the time-out procedure TOP before start of surgery importantly add to safety of the entire perioperative process.

Invasive monitoring is recommended in open procedures with expected severe blood loss. Invasive monitoring involves the use of a central venous line and an arterial line next to conventional monitoring capnography, ECG, pulse oxymeter, FiO2, temperature, and urine output and precordial Doppler. The major goals of invasive monitoring are timely recognition of serious problems that may occur in open craniosynostosis surgery, such as hypohydration, hypotension, and electrolyte disturbances, as well as being able to check effectiveness of treatment of these problems.

Venous cerebral congestion by hyperflexion of rotation of the head must be avoided. Usually, the patient is placed in a moderate anti-Trendelenburg position to reduce blood loss cave air embolism, see below under Complications. Notably, in children with exorbitism the eyes should be well protected with eye cream, and pressure on the eyes must be avoided especially in prone position. Single administration of an antibiotic, after the induction, before start of surgery is standard procedure.

Most recommendations are aimed at a balanced technique that provides for cardiovascular stability with the use of opioids and volatile agents next to relaxants. The use of remifentanil infusion 0.

An additional problem is that measurement of the lost volume intraoperatively is impeded by surgical technique and type of patient. Few studies have been performed to optimize blood loss measurement and findings are mostly not conclusive. Infiltration of the skin with vasoconstrictors before the incision is much used but it's effectiveness is disputed because of the greater degree of bleeding with treatment of the peristium and bone.

Controlled hypotension is rarely used because it has no clear benefits and can be disadvantageous in case of ICP and also in the anti-Trendelenburg position. The acute normovolemic hemodilution ANH technique is used before the surgical intervention after the child is brought under anesthesia.

It involves removal of whole blood via the arterial or central line which is replaced with colloids or with crystalloids according to precise calculations and formulas. According to a B study, this technique in itself is not sufficient to reduce or avoid homologous blood transfusion in this type of surgery.

Duncan 82 found no difference in allogeneic transfusion rate when a cell saver was used and when not. Intraoperative antifibrinolytics. Aprotinin: is no longer used after serious complications in cardiac surgery. Tranexamic acid: its use in itself is controversial, but is has been applied in combination with other methods.
Guidelines for the application of blood products are in place in every hospital as established by the local transfusion committees, and in conformity with the CBO Guideline Blood transfusion Venous air embolism VAE and the subsequent cardiovascular collapse can be prevented by a precise technique and by rapid application of adequate monitoring precordial Doppler, capnography, echocardiography, transcutaneous O₂—CO₂ monitoring, esophageal stethoscope, and venous central line.

Although Faberowski 86 reports an incidence of Meyer 88 reports a 2. The hemodynamic consequences of VAE are generally insubstantial, provided the patients are well monitored and measures are taken upon signs of VAE.

As a result a massive blood loss the patient may develop relevant consumption coagulopathy and dilutional coagulopathy characterized, in principle, by depletion of soluble clotting factors. The situation changes when these children undergo a distraction osteotomy procedure and need to be intubated acutely in case of respiratory insufficiency, or when the distraction materials are being removed.

Evidence from the C level studies is conflicting on this issue. One of the 2 studies recommends immediate fiberoptic intubation, whereas the other study claims that the distractors exert minimal effect in the anesthesiologic conditions when certain factors must be taken into account the right screwdrivers and cutting pliers must always be available and that removal of the vertical bar allows for direct laryngoscopy.

The possibility of cerebral salt wasting syndrome should be considered when a patient develops hyponatremia after craniosynostosis surgery. Criteria for extubation at the end of the procedure, before transport to the ICU, are the following: rapid recovery of spontaneous and stable breathing, hemodynamic stability, normothermia, short to medium operation time with relatively little blood loss, and no continuous large blood loss through the surgical drains.

In principle all studies recommend ICU admission after open craniosynostosis surgery to continue volume management there and, if necessary, start artificial respiration. Most studies on postoperative transfusion management report a tendency to overtransfusion.

There is scarce literature on postoperative pain treatment after craniosynostosis surgery. In Rotterdam 2 double blind, placebo-controlled studies have investigated pain treatment in the first 24 hours after cranial correction for craniosynostosis. Van der Marel 95 treated 20 children with oral paracetamol and 20 with rectal paracetamol. The pain scores in the oral group were higher, but this effect disappeared with exclusion of the patients who had vomited after oral administration.

Despite the fact that Rectal administration of paracetamol was recommended. Prins 96 describes 12 children who received intravenous paracetamol and 14 children who received rectal paracetamol. This is indicative of more discomfort, possibly caused by more pain, and intravenous administration was recommended. In the European literature, the use of paracetamol and NSAIDs is recommended if the blood loss is not significant with or without codeine phosphate British literature.

All textbooks emphasize, and this also appears from our own practice, is that the pain scores are surprisingly low. In patients operated on for nonsyndromic craniosynostosis, invasive monitoring can usually be discontinued the following day.

Despite the proven effectiveness of erythropoietin and blood drawing for autologous transfusion, the use of these strategies is discouraged as they involve high costs and require repeated venipuncture, which is not child friendly. Introduction of less invasive interventions is associated with less blood loss. As these, however, are very young children with a smaller circulating capacity, blood loss relatively is still significant. In selected cases admission to a medium care unit could be considered defined as a monitored bed providing for artificial respiration.

The anesthesiologic goals in craniofacial surgery can only be realized in specialized pediatric centers, where multidisciplinary perioperative care is provided by a team composed of plastic surgeon, neurosurgeon, maxillofacial surgeon, pediatrician, pediatric anesthesiologist, pediatric intensivist, and specialized pediatric nurses with experience and means to manage and monitor this type of patients and where a sufficient number of children is operated on to keep the experience of the team at a high level.

Administration of EPO preceding the intervention, as well as collecting autologous blood for autotransfusion are advised against. Postoperatively a bed in a PICU must be available. In less drastic interventions medium care may perhaps suffice, although 1 should be aware that these are mostly very young children with a smaller circulating capacity, in whom even slight postoperative blood loss must be monitored carefully.

Invasive monitoring is recommended in the case of open procedures with expected severe bleeding. Overtransfusion in the postoperative phase should be prevented by adhering to the guideline on transfusion management. Extubation at the end of the operation, before transport to the ICU, is possible in case of rapid recovery of spontaneous and stable breathing, hemodynamic stability, normothermia, short to medium operation time with relatively little blood loss, and no continuous large blood loss through the surgical drains.

The analgesic of choice for postoperative pain treatment is paracetamol. If this is not sufficient, an NSAID can be added even though this could increase the risk of postoperative bleeding.

What are the indications for surgery in the different types of nonsyndromic craniosynostosis? What treatment is most indicated for the different types of nonsyndromic craniosynostosis? Nonsyndromic craniosynostosis can present in varying severity of the deformity of the skull. Surgical treatment seems indicated on the basis of: The variability in both the severity of the morphologic abnormality and in the occurrence of increased ICP may occasionally result in less strict operative indication on the basis of these parameters.

Many different surgical techniques have been described for the treatment of nonsyndromic craniosynostosis but the functional results of these techniques have not always been shown convincingly. In addition, the timing of surgery is debated. For patients with a synostosis of multiple cranial sutures in whom a syndromic diagnosis cannot yet be made, the method of treatment of syndromic craniosynostosis is recommended.
This chapter will place a focus on craniosynostosis of the sagittal suture scaphocephaly, metopic suture trigonocephaly, unilateral coronal suture frontal plagiocephaly, and lambdoidal suture pachycephaly. The indications for treatment of craniosynostosis are the risk of ICP and the morphologic abnormality of the skull and face. The chapter on cognition and behavior provides evidence of absence of a relation between cognitive functions in children with a nonsyndromic craniosynostosis and surgical treatment or not, severity of the deformity, or age at operation.

The prevention or treatment of the associated neuropathology is therefore no indication for surgery. The morphology can vary from very mild to an evidently abnormal skull shape. In sagittal suture synostosis, compensatory growth of the other cranial sutures results in increasing deformity in the course of time, so that, for example, the frontal bossing in scaphocephaly can intensify after birth. Barritt evaluated a series of 44 children with scaphocephaly, of whom 34 did not undergo surgery.

This was common policy in this clinic at the time for children presenting with this condition at an age older than 6 months. In these 34 children, the deformity in any patient did not seem to improve in the first 10 years. The presentation of trigonocephaly also varies from very mild to severe.

Its mildest form is characterized only by a bony ridge as a result of premature fusion of the metopic suture. In follow-up, cosmetic worsening is only rarely seen. The other end of the spectrum is characterized by a wedge-shaped forehead with hypotelorism and supraorbital retrusion on the basis of decreased growth of the forehead widthwise. Frontal plagiocephaly also shows some variation in phenotype, notably in facial asymmetry.

Unilateral synostosis of a lambdoidal suture causes increasing asymmetry of the back of the head and particularly also of the face. Increased ICP does not seem to be prominent in this type of synostosis, although reliable research on this issue is lacking see chapter 8. Since the first operative intervention for craniosynostosis, many surgical techniques for the various types of craniosynostosis have been described.

A broad distinction is made between osteoclastic techniques and remodeling techniques. In the first group bone is removed, enabling the developing and expanding brain to change the shape of the skull, in part because the adverse impulse to the growth direction of the skull also is removed.

From this technique evolved the remodeling techniques, because it was suspected that one cannot only rely on the self-correcting capability of the skull and the brain. These techniques are aimed at directly achieving the desired skull shape by a kind of reconstruction.

Randomized comparisons of the different surgical techniques have never been performed. For the different types of nonsyndromic craniosynostosis we will provide a review of the literature regarding the different techniques in these 2 groups, but also pay attention to the new developments in the past 10 years in a separate section. Ingraham described a technique using bilateral parasagittal strip craniectomies, in which the fused suture was not resected.

To prevent the problem of premature reossification of the skull bones he applied a layer of plastic to the bone rims. Successful treatment required early intervention, however, preferably in the first 2 months of life. The results of this technique were confirmed by other authors. Albright presented a sagittal strip craniectomy with biparietal wedge osteotomies. The same procedure was performed by Marucci in 89 patients, but they noticed that 7 of the 89 patients later developed a cosmetically unsatisfactory vertex bulge, which they interpreted as a sign of possibly raised ICP.

Also Florisson reported that some patients with scaphocephaly again showed signs of raised ICP postoperatively. Amm presented a technique, in which the usual 2. They explicitly try to make use of gravitational force by keeping the infant strictly in supine position during sleep for a period of 3 weeks.

Using this technique an average improvement of the CI of 8. All in all, many of the described techniques represent only slight modifications of previously presented techniques and not real conceptual changes. The sagittal suture itself is not resected, and the temporoparietal bone is bent outwards. In this technique, the anteroposterior diameter of the skull is dynamically shortened by attaching the bone strip in the middle of the skull, which is shortened, to the frontal bone.

Thus, it is in fact also a compression technique, which might be associated with an increase in ICP. Only a global development quotient, however, was tested in very young patients, without inclusion of a control group, which raises great doubt on the reliability of this finding. In older children or in children with a more pronounced dysmorphology notably frontal bossing, a more extensive calvarial reconstruction is needed, with resection and reconstruction of the frontoparietal complex.

The authors concluded that in this population this more extensive procedure had no adverse effect on postoperative growth rate of the skull or on intracranial volume. Lauritzen reported in on the first operations in which distraction was used to correct craniosynostosis. This method is eminently suitable for scaphocephaly in view of the unidirectional growth delay widthwise. The results are not worse, but also not better than those of the current techniques and the small numbers preclude valid comparison.

Also regarding the newer surgical techniques, randomized or comparative studies have not been published. Also in view of the complex three-dimensional dysmorphology seen in metopic synostosis, treatment by suturoectomy solely is considered insufficient. Posnick analyzed the growth process on the basis of CT data in 10 patients and concluded that correction of the hypotelorism had remained insufficient.

Selber found that the use of interpositional bone grafts in a series of 68 metopic children led to a decrease in the development of temporal dents. In a population of 92 trigonocephaly children in which no bone graft was used, Van der Meulen, however, observed an automatic growth correction of the hypotelorism on x-ray, on account of which the use of an interpositional bone graft was judged to be unnecessary.

From a photographic evaluation of 45 patients, Hilling concluded that a satisfactory esthetic outcome on the long term was largely dependent on a good initial reconstruction. A distinction is made between unilateral and bilateral frontal advancement techniques.
Some surgeons prefer the unilateral advancement technique so as to be able use the unaffected side as a reference for the degree of advancement, whereas others maintain that adequate advancement with good symmetry is only possible if the entire frontal bone and both orbital margins have been dissected.

Barone and Jimenez report since the late s on endoscopic strip craniectomy of the affected suture. The benefits mainly lie in low morbidity, short hospitalization, and little need of blood transfusions in comparison with the classic fronto-orbital advancement technique. The cosmetic outcome is reported as good, but it not adequately quantified. For this reason, the technique was adapted to an endoscopic unilateral fronto-orbital advancement technique. This was applied in 10 patients, who did not develop any complications, did not need blood transfusions, and could be discharged home after 2 days.

The cosmetic outcome was assessed to be similar to that of fronto-orbital advancement. The number of publications on lambdoid suture synostosis is very limited, probably because of the very low prevalence. Unilateral lambdoid suture synostosis causes only relatively little volumetric restriction. The morphologic changes consist of an asymmetry of the face and an asymmetric basal occipital region.

Kim reports a study comparing distraction versus remodeling surgery. Among the patients were 3 children with brachycephaly treated with the distraction method and 4 with remodeling. The distraction method led to satisfactory calvarial expansion and an esthetically pleasing outcome. Advantages of distraction are the significantly shorter operation time, less bleeding, shorter stay at the neonatal intensive care unit NICU, and better skull shape.

A disadvantage of the distraction method is that children must have reached the age of 6 months. The authors are of the opinion that these downsides do not outweigh the benefits of distraction. Most children with a craniosynostosis are operated on in the first year of life. A direct evaluation of surgical outcomes is possible, but a final evaluation is not possible until the child has fully grown at adult age.

As a surgeon's career spans perhaps 25 year, and at most 30 years, the surgeon can only at an advance career stage assess the result of his or her own work.

There may be a tendency to ascribe a poor result to a wrongly performed surgical technique, without questioning the technique itself. Furthermore, knowledge about the natural course of the condition with and without surgical intervention is usually lacking. This shortcoming undoubtedly is corespansible for the fact that we see regular shifts in type of intervention in the course of decades and that paradigms change.

One and the same research group may recommend a certain approach in the 1 decade and disassociate oneself from it in the next decade. Only few solid objective results with such a long follow-up are available, whereas comparative studies of different surgical techniques with or without randomization have not at all been performed.

Moreover, notably the larger series yield mixed data, because all craniosynostoses, including the syndromic types, are pooled. One way to assess the outcome of surgery is looking at the operative morbidity and mortality. Most series are too small, however, to yield significant data.

Morbidity is reflected in the complications that may occur and length of hospitalization. Although the first is a relatively objective score, the second is certainly not. Length of hospitalization strongly depends on the treating physicians, but also on the healthcare system itself. To illustrate this, for similar procedures the average length of hospitalization in North America is significantly shorter than in Europe.

The patients are simply discharged home earlier, which possibly is enabled by a better outpatient care system. Generally, hospitalization after minimally invasive interventions is 1 or 2 days; for more complex interventions 2 to 7 days. As postoperative infections are very rare, only large series can yield significant data.

In series of patients who underwent surgery for unicisutural craniosynostosis only 2.0. In general, the complications rates in the nonsyndromic craniosynostoses are significantly lower than in the syndromic craniosynostoses, 3.

The use of metal plates and screws has been customary in a certain period. Meanwhile, it has become evident that application of this material in young children may lead to intracranial translocation of the plates and screws, with the screw ends penetrating the dura.

In the past few years, resorbable plates and screw devices have been applied for specific indications in craniosynostosis surgery. The outcome of surgery can also be expressed by need for secondary revision surgery. This is rarely needed. Amm routinely uses three-dimensional surface analysis software to analyze CT-scans.

Computerized tomography scanning, however, is a source of radiation exposure. Still it enables to assess the outcome in a relatively reliable and objective manner, and this yielded a mean 8. Marucci noticed a cosmetically unsatisfactory vertex bulge after minor surgery in 7 of 89 patients. At follow-up in the course of years after reconstruction of trigonocephaly, the lateral sides of the forehead often appear to revert to a certain degree and temporal depressions may be seen, which however seldom are cosmetically unsatisfactory to the degree that revision surgery is justified.

Hilling evaluated the result of bifronto-orbital advancement in trigonocephaly from photographs of the patients pre- and postoperatively and after follow-up by a panel of independent assessors with the aid of a scoring form.

They concluded that the cosmetic result generally was good to very good, but also that the severity of the deformity does not influence the eventual outcome. In addition, they established that the obtained result is stable during time and independent of the age at operation 6 to 15 months. Temporal depressions are ultimately the most frequent abnormalities at follow-up and cosmetically usually only little unfavorable.

Van der Meulen performed standard x-rays of the skull before and after operation and in the course of time and noted that although in fronto-
Timing of the first cranial vault expansion may differ; it is often performed by protocol when the child reaches a certain age, but in some centers not international centers either occipital expansion or fronto-orbital advancement is performed as first cranial vault expansion. In case of orbital involvement, correction should include remodeling of the supraorbital margin ie, trigonocephaly, frontal plagiocephaly, and abnormality.

Cranial remodeling in nonsyndromic craniosynostosis should take place in the first year of life. Strip craniectomy for nonsyndromic craniosynostosis is in principle not indicated for mild types of craniosynostosis, such as the metopic ridge and the partial synostosis of the sagittal suture, with few morphologic abnormalities. In all other types, operative correction of the deformity is indicated both on functional and cosmetic grounds.

Although the risk of relapse seems to be somewhat increased after an early limited intervention, the risk is probably small. This should be weighed against the burden of a late complete cranial remodeling with longer operation time and more bleeding. This guideline therefore does not pronounce on a preference for early or late surgery. Still, in view of the results of Renier regarding the outcome of surgery after the first year of life, it is recommended to perform the intervention in the first year of life.

The earlier mentioned panel assessment of photographs by Hilling in trigonocephaly, was also applied by the same researchers in 59 patients with frontal plagiocephaly. Also in this deformity, the temporal depression is the most striking finding at follow-up. Direct evidence for this supposition, however, is lacking. For that matter, this research group also concludes that the frequency of temporal depressions decreases with increasing experience of the surgeon.

Lo specifically assessed the orbital morphology both of bone and soft tissue after surgical intervention and evaluated this by means of CT-scans. They noted that the orbital morphology had genuinely improved, but also that complete symmetry had not been obtained. Furthermore, outgrowth, however, was normalized. The timing of surgery differs worldwide and is dependent, among other things, on the surgical technique used.

In patients with boat-shaped head, it was observed that the initially improved CI index had slightly decreased 1 year after correction, whereas in fronto-orbital advancements, the frontolateral skull will again slightly recede a part reversal to the primary deformity. This in general does not cause cosmetic problems, the more so because some surgeons in anticipation will apply overcorrection.

Thompson proposes that nonsyndromic craniosynostosis ideally is corrected within the first year of life, with scaphocephaly as a possible exception, corrected with early modified strip craniectomy. The largest series is reported by Marchac, nearly patients with all types of craniosynostoses were operated on in Paris. It is suggested to correct brachycephaly at the age of 2 to 4 months often syndromic patients with raised ICP and the other conditions at the age of 6 to 12 months.

The reason why is not argued, however. Amm prefers correction of scaphocephaly at the age of 6 to12 months, without clear argumentation. Fearon, evaluating 16 children with trigonocephaly, established that the younger the child, the more spontaneous improvement of the hypotelorism after surgery, even though the hypotelorism itself was not surgically corrected. In , they concluded that surgery at the age of 3 months would be ideal for good development of the orbits.

Thirteen years later, however, the same research group concludes that the younger the child, the larger the growth inhibition of the skull after operative intervention. Their timing then is 4 months for the scaphocephalic child to prevent progression of the frontal bossing and 9 months for monosutural craniosynostoses requiring advancement so that in any case the frontal band is strong enough, but the dural capacity of bone regeneration also is still sufficient to close all holes.

Also for frontal plagiocephaly, there is no consensus on the timing of surgery. Although minimal invasive surgical techniques need to be performed at young age, ages of 6 months to from 15 to 18 months are recommended for major reconstruction. Here, too, the most convincing argument for late surgery at the age of 18 months is a possibly lower chance of later relapse.

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The novel techniques, such as the endoscopic strip craniectomy and the spring-assisted craniectomy, seem safe and effective. There is some concern in the field about the use of the remodeling helmet after endoscopic strip craniectomy. In patients with a NSOP this helmet exerts a restrictive influence on skull growth.

This is notably an undesirable effect in craniosynostosis. It would be prudent, therefore, to systematically monitor patients treated in this way on restricted skull growth and signs of raised intracranial pressure.

Considering the findings from the study of Marucci, this holds true as much for patients treated with extended strip craniectomy. Surgical correction is in principle not indicated for mild types of craniosynostosis, such as the metopic ridge and the partial synostosis of the sagittal suture, with few morphologic abnormalities. In all other types, operative correction of the deformity is indicated both on functional and cosmetic grounds.

Cranial remodeling in nonsyndromic craniosynostosis should take place in the first year of life. Strip craniectomy for nonsyndromic craniosynostosis without additional measures is advised against. The choice between the other techniques for scaphocephaly ie, extended strip craniectomy, complete cranial remodeling, and strip craniectomy combined with helmet or springs is based on age of presentation and severity of the abnormality.

In case of orbital involvement, correction should include remodeling of the supraorbital margin ie, trigonocephaly, frontal plagiocephaly, and brachycephaly. The use of metal plates and screw devices in cranial remodeling in very young children is strongly discouraged. What type of cranial vault expansion is at what moment indicated as first treatment of the various types of syndromic craniosynostoses? In the different international centers either occipital expansion or fronto-orbital advancement is performed as first cranial vault expansion.

Timing of the first cranial vault expansion may differ; it is often performed by protocol when the child reaches a certain age, but in some centers not
The group of complex craniosynostoses clinical suspicion of syndromic craniosynostosis without proven genetic abnormality, often with multiple sutureal synostosis is managed in the same way as the syndromic craniosynostoses, as these patients often experience similar problems regarding cognition and increased ICP.

In the mids, a number of articles were published reporting the experiences of various large centers, in which in particular the frontal cranial vault expansion was propagated. At later referral, a more extensive fronto-orbital advancement was performed using tongue in groove. McCarthy et al described a comparable procedure in a group of 76 patients with a craniofacial syndrome.

All underwent a primary fronto-orbital advancement within 18 months mean 6. The authors propose fronto-orbital advancement by protocol at the age of 6 to 9 months. In more than half of these patients the indication was increased ICP. In 1 patient a third cranial vault expansion was performed. The benefits of a primary frontal expansion for a young child were considered too short lasting; it would have only a minimal effect on the remainder of the facial deformity and a next intervention for a facial correction Le Fort III, monobloc or facial bipartition would therefore be unnecessarily compromised.

Exception: severe exorbitism posing a threat to vision, for which fronto-orbital advancement should indeed be the first procedure. The classic management includes initial anterior skull remodeling and facial advancement as a second step. Since evolution of these complex facial craniosynostoses results in turricephalic aspect or recurrences more often than nonsyndromic craniosynostosis, another approach can be used, consisting in an initial posterior vault expansion.

Honnebier reports that nearly half of the patients with Muenke syndrome required a second fronto-orbital correction to correct retrusion of the supraorbital margin following an initial fronto-orbital advancement at the age of 6 to 9 months. The authors do not provide a recommendation to adjust the timing of the initial intracranial intervention to this result. Fronto-orbital advancement is recommended for the Muenke and Saethre-Chotzen syndromes, at 9 to 12 months for Muenke syndrome and at 6 to 12 months for Saethre-Chotzen syndrome.

The difference in timing is based on the low risk of increased ICP in Muenke syndrome. The team from Dallas provides arguments for parietal widening at very young age 8 to 16 weeks in severe presentation of the Pfeiffer syndrome. Occipital correction is not performed because they prefer to combine this with expansion of the foramen magnum in case of a symptomatic Chiari.

For the milder types, they perform fronto-orbital advancement at the age of 9 to 15 months. Their strategy seems to be based on setting a low threshold for placement of a tracheostoma and not considering a monobloc procedure until the age of 3 or 4 years in such a situation.

Jeevan in issued a warning relating to this intervention: cave abnormal transosseous venous drainage! Fronto-orbital advancement is recommended for the syndromic types without midface hypoplasia. Serious complications in 36 monoblocs with distraction were death by acute tonsillar herniation postoperatively in 1 patient and septic osteonecrosis of the frontal bone in 1 patient.

Earlier operations preceding the monobloc increased the morbidity. Parisian studies on mental development in syndromic craniosynostosis patients report higher IQ in patients operated on in the first year of life. This was found for patients with Apert syndrome, Crouzon syndrome, and Muenke syndrome with bicoronal synostosis, but not for patients with Muenke syndrome with a unilateral coronal suture synostosis see chapter From analysis of the treatment protocols of the craniofacial centers participating in the International Society for Craniofacial Surgery it appeared there was consensus to perform the initial cranial vault expansion within the first year of life.

The London team is the only 1 with a different approach. Cranial modeling is performed as soon as the VEPs show signs of delays in optic nerve conduction. Of the 24 Apert patients treated according to this protocol, 20 developed increased ICP. Metal plates and screw devices have been commonly used during a certain period.

Later it became evident that application of this material in young children may lead to intracranial translocation of the plates and screws, with the screw ends penetrating the dura.

In the past few years resorbable plates and screw devices have been applied for specific indications in craniosynostosis surgery. It is believed that occipital cranial vault expansion results in a larger intracranial volume than does a fronto-orbital advancement. Leaving the fronto-orbital region untouched in the first operation leads to lower risk of complications of a monobloc procedure performed at a later stage.

The question arises whether intensive ophthalmologic screening for increased ICP as well as the practice of performing cranial vault expansion not until results are abnormal should be protocolized.

The high frequency of hospital visits and tests places a burden on patients, parents, and the members of the craniofacial team. In addition, there is a risk of false-negative VEP results, which does not seem to outweigh the lower number of patients who will not be operated on for the indication of increased ICP. The initial cranial vault expansion in syndromic craniosynostosis is performed within the first year of life.

In patients with Muenke syndrome is recommended to perform the operation between the ages of 9 and 12 months. The use of metal plates and screw devices in very young children is strongly discouraged. What treatment is most indicated for maxillary hypoplasia sagittal, vertical and transversal, including exorbitism? The focus of this document is on maxillary hypoplasia in syndromic craniosynostosis patients; the exorbitism is a consequence of this hypoplasia and is therefore not separately investigated.

Furthermore, correction of hypertelorism is evaluated as part of the monobloc operation. The Apert, Crouzon, and Pfeiffer syndromes are associated with hypoplasia of the maxilla, exorbitism, and hypertelorism. Various techniques are available to correct these deformities, and their...
timing greatly influences the eventual outcome. Corrections of the sagittal and vertical dimensions are discussed first, followed by those of the transversal dimensions.

As investigated by Posnick in, the face grows in 2 distinct periods. Craniofacial growth in the first 6 or 7 years is determined by growth of the brain, eyes, and the nasal cartilages. This observation is likely to be important for the timing of corrective surgery.

Although some authors suggest that further growth could be restricted by surgery, others report very slow unchanged growth after surgery in syndromic craniosynostosis patients see below.

Bachmayer determined growth of the midface in 19 surgically treated syndromic craniosynostosis patients Apert, Crouzon, and Pfeiffer syndrome aged from 6 to 15 years. Postoperative growth of the maxilla in sagittal direction was less than 0. In vertical direction, the growth was 1. Meazzini concluded that the sagittal growth in untreated syndromic craniosynostosis patients was negligible, and if minimally present would not be adversely influenced by an operation.

Kreiborg analyzed 8 patients with Crouzon or Apert syndrome on preoperative growth, stability of the Le Fort III osteotomy, and postoperative growth. Neither group showed postoperative horizontal and vertical growth. Fearon states that the disturbed maxillary growth in the syndromic craniosynostosis patients is intrinsically associated with the syndrome, and that this would not so much be the effect of an operation.

Covers bowed. This copy does have a cigarette smoke odor. First Edition. Garrison-Morton The article begins: "Dr Robert Langston was training as a general surgeon in Scotland in when a chance encounter with the imperious Sir Harold Gillies led him to work in the famous Rookwood burn unit near London.

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How to optimize recognition of craniosynostosis in the primary and secondary healthcare sectors? Craniosynostosis should be recognized timely and cognition, and problems with school choice. Psychosocial problems, such as having to cope with negative reactions from others, a possible discrepancy between deviating physical appearance and the syndromic craniosynostoses, and in patients with Apert syndrome always present in a severe form.

In the syndromic group, abnormalities such as astigmatism and strabismus are very frequent. Deformities of the extremities are notably restricted to malformation, ventriculomegaly, and hydrocephalus. In all types of syndromic synostosis hearing loss is described. Vision, refraction and motility problems of cognition and ICP. Associated brain abnormalities in syndromic craniosynostosis include raised intracranial pressure ICP, Chiari I malformation, ventriculomegaly, and hydrocephalus. In all types of syndromic synostosis hearing loss is described. Vision, refraction and motility abnormalities in the nonsyndromic craniosynostoses are in fact only seen in unilateral coronal suture synostosis.

The group of complex craniosynostoses is managed in the same way as the syndromic craniosynostoses, as these patients often experience similar morphologic abnormalities of the cranium, the orbits, and the upper jaw.

Treatment has a 2-fold aim: to enlarge the cranial volume so as to prevent sequelae of ICP mental retardation and impaired vision, and to correct the skull shape by a reconstruction. The surgical treatment of patients with syndromic craniosynostosis was developed in Paris in the early s.

Through crisp line drawings of the face, lacrimal system, and orbit, each chapter systematically outlines the relationship of physical structure to the procedure. Plus, you'll find step-by-step surgical approaches for functional and cosmetic problems, along with the latest advances in lacrimal surgery, ptosis repair, blepharoplasty, and more! Key features: - Anatomy introducing every chapter, providing you with the tools to completely understand and map out an operative approach - A practical, systematic orientation moving from external to internal structures - Nearly crystal clear illustrations that depict anatomy and techniques - The expertise of prominent physicians who offer a wide scope of diagnostic and treatment insights that can be tailored to your own situation.

Second Edition: First Printing. Boards and pages are clean, unmarked, brightly colored, tightly bound and sharp cornered but has slight scuffing at top of spine from shelf wear. Book is in English, translated from the original German. Contains illustrations.

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Consequently, displacement of the ear and lateral deviation of the nose occur. Bilateral synostosis of the coronal sutures result in retraction of the forehead and the supraorbital rim with a broad skull. Syndromic craniosynostosis is usually characterized by synostosis of both coronal sutures, but other types of craniosynostosis are seen as well. Indications for surgical treatment of nonsyndromic craniosynostosis are the risk of increased intracranial pressure ICP and the abnormal skull shape.

Since the first surgical intervention for craniosynostosis, a great many surgical techniques for the various types of craniosynostosis have been described. A broad distinction is that between osteoelastic techniques and remodeling techniques, osteoelastic techniques involve resection of bone, thereby allowing the developing and expanding brain to change the shape of the skull, also because the adverse impulse to the growth direction of the skull is removed.

The remodeling techniques, on the other hand, do not rely on the selfcorrecting capacity of the skull and the brain, but attempt to obtain the desired skull shape by a reconstruction. The surgical treatment of patients with syndromic craniosynostosis was developed in Paris in the early s.

The group of complex craniosynostoses is managed in the same way as the syndromic craniosynostoses, as these patients often experience similar problems of cognition and ICP. Associated brain abnormalities in syndromic craniosynostosis include raised intracranial pressure ICP, Chiari I malformation, ventriculomegaly, and hydrocephalus. In all types of syndromic synostosis hearing loss is described. Vision, refraction and motility abnormalities in the nonsyndromic craniosynostoses are in fact only seen in unilateral coronal suture synostosis.

In the syndromic group, abnormalities such as astigmatism and strabismus are very frequent. Deformities of the extremities are notably restricted to the syndromic craniosynostoses, and in patients with Apert syndrome always present in a severe form.

In the other syndromic types, deformities of the extremities are generally very mild. Parents are often confronted with health professionals who do not recognize the craniosynostosis shortly after the birth of the child.

This can be a source of stress for parents and lead to a delay in diagnosis and treatment. Nonsyndromic and syndromic craniosynostosis may co-occur with cognitive impairments and behavioral disorders. These may occur both intrinsic to the congenital defect and secondary to, for example, increased cranial pressure or abnormal physical appearance. Notably, families of a child with syndromic craniosynostosis may experience psychosocial problems, such as having to cope with negative reactions from others, a possible discrepancy between deviating physical appearance and cognition, and problems with school choice.

How to optimize recognition of craniosynostosis in the primary and secondary healthcare sectors? Craniosynostosis should be recognized timely
It appears, however, that craniosynostosis patients often are not referred at all or referred too late. A complicating factor in the recognition of craniosynostosis is the high incidence of positional head shape deformities. It may be expected that recognition, and thus referral of patients with craniosynostosis will happen earlier if health professionals in the primary and secondary healthcare sectors are facilitated to make this distinction.

Preceding the referral to a tertiary healthcare center, we often see overuse of diagnostic imaging, which is associated with further delay in referral, an extra burden and insecurity for patient and parents as well as unnecessary expenditure.

This should be restricted to a minimum. There are many options to diagnose isolated craniosynostosis or any associated abnormalities. To restrict both underdiagnosis and overdiagnosis, basic question 3 concerns further diagnostic procedures in a tertiary healthcare center. Areas of attention are diagnostic imaging, genetic diagnostics, and the role of the pediatrician. Inclusion of a clinical geneticist in a multidisciplinary craniofacial team provides the opportunity to address questions from parents and attending physicians.

For the attending physician it is important to know if there are any associated anomalies to be expected next to the craniosynostosis that may be of influence to the treatment plan and the prognosis of the child. The abnormal skull shape is recognized by parents themselves, midwife, obstetrician, general practitioner, infant health clinic physician, nurse, pediatric physiotherapist, pediatrician, pediatric neurologist, and helmet maker.

Occasionally, it is recognized later by other specialists and sometimes not until a craniofacial team is involved. These data are from the Birmingham craniofacial center and could therefore well compare with the situation in the Netherlands. Of the 89 referrals with the initial diagnosis nonsynostotic occipital plagioceplahy NSOP made by the referrer, 10 patients appeared to have a craniosynostosis. A craniosynostosis or positional skull deformity is primarily recognized by physical examination, notably skull shape 4, 6 in combination with history taking, 3, 5 and an imaging study is rarely indicated.

This deserves great attention because early recognition and surgical correction are essential to a satisfactory treatment outcome. Ridgeway recommends a number of diagnostic steps that should lead to early diagnosis, in which history taking and physical examination yield the most important information to distinguish between positional head shape deformities and craniosynostosis.

Using this information in a flowchart at intake appears to be a safe method to make this distinction at the earliest. This will prevent delay in start of treatment as a possible consequence of the increase of patients of positional head shape deformities. There are no other publications on referral patterns in children with abnormal skull shape.

History taking should address the distinction between positional head shape deformities and craniosynostosis on the one hand, and risk factors and associated anomalies on the other hand. Craniosynostosis is present at birth, positional head shape deformities usually not. Presence or absence of a preferred sleep position is essential in the diagnostics of positional head shape deformities.

Analysis of a flowchart to differentiate positional head shape deformity versus craniosynostosis by means of a questionnaire completed by telephone or e-mail showed that the questionnaire recognized no more than More in detail the different craniosynostoses can be recognized as follows: The anterior fontanel shows earlier fusion or is triangular shaped rather than diamond shaped in synostosis of 1 adjacent suture. Mid-face hypoplasia, proptosis, and hypertelorism are found particularly in the syndromic types of craniosynostosis.

The following characteristics may help to distinguish craniosynostosis from positional head shape deformities: American Cleft Palate-Craniofacial Association ACPA recommends that craniofacial teams provide education to health professionals in obstetrics and neonatology as well as GPs and infant health clinic physicians so as to improve early recognition of children with craniofacial deformities.

Grol emphasizes that effective implementation of guidelines requires behavioral changes in physicians. E-mail consultations improve the quality of decision-making, notably as they are easily accessible. Audit programs are most effective when integrated with other educational and research programs. Often it takes long before parents and the child with craniosynostosis are referred to the tertiary center, which may lead to treatment delay and confusion, and consequently a great deal of parental stress.

The flow diagram to discriminate between positional skull deformities and craniosynostosis was validated in a tertiary care setting, where it was applied by secretaries who were not specifically trained in distinguishing between these 2 conditions.

Consequently, we do not expect great differences in its reliability when applied in primary and secondary care settings. Ideally, validation for primary and secondary care settings finds place in these settings, but this does not seem to be practically feasible in view of the very low frequency of craniosynostosis compared with positional skull deformities. Additional imaging studies to differentiate between positional skull deformities and craniosynostosis are rarely performed in tertiary centers.

On clinical diagnosis of a positional skull deformity, skull x-ray or ultrasound are advised against to prevent unnecessary medical imaging with associated costs, radiation exposure, burden for patient and parents, lack of added value, and required experience in interpretation.

Consultation with a tertiary craniofacial center is indicated in suspected patients, for which submitting normal pictures front view, lateral view, anterior view, and view from above will usually suffice. The following measures are essential to optimize recognition of craniosynostosis in the primary and secondary care sectors: Provide structured education and training instruction to infant health center physicians, GPs, midwives and obstetricians about over skull deformities via centers of expertise with an initiating role for the tertiary centers.

Provide feedback about the referral pattern on the basis of an analysis of national registry data focus on patient's age at referral to tertiary center.
The adequate performance and interpreting of dysmorphologic examination is one of the specific medical competencies of a clinical geneticist. Data

The January issue of the American Journal of Medical Genetics Part A is fully devoted to dysmorphologic examination of the face and extremities.

On indication, dysmorphologic examination of the whole patient is repeated. Possible indications are the development of new physical problems, measurements are made and compared with reference values.

Distinguish between nonsyndromic and syndromic patients on the basis of the presence of dysmorphic characteristics. In addition, several bodily diagnostiscs. Craniosynostosis is a birth defect that can present both in isolated form and syndromic form.

An etiologic diagnosis also provides for determining the risk of recurrence as well as the alternative choices in new pregnancies, such as prenatal diagnostics. Craniosynostosis is a birth defect that can present both in isolated form and syndromic form. An etiologic or classifying diagnosis allows for making a prognosis not so much regarding the craniosynostosis, but rather regarding the child's general and psychomotor development and possible associated anomalies.

An etiologic diagnosis also provides for determining the risk of recurrence as well as the alternative choices in new pregnancies, such as prenatal diagnostics. Craniosynostosis is a birth defect that can present both in isolated form and syndromic form.

In line with the international literature, we distinguish between nonsyndromic and syndromic craniosynostosis. Dysmorphologic examination may distinguish between nonsyndromic and syndromic patients on the basis of the presence of dysmorphic characteristics. In addition, several bodily measurements are made and compared with reference values.

On indication, dysmorphologic examination of the whole patient is repeated. Possible indications are the development of new physical problems and newly observed delay in development in the course of time.

The January issue of the American Journal of Medical Genetics Part A is fully devoted to dysmorphologic examination of the face and extremities. The adequate performance and interpreting of dysmorphologic examination is one of the specific medical competencies of a clinical geneticist. Data
of patients referred from through and of patients referred from through were compared on relative incidence of occurrence of the various types of craniosynostoses.

The total number of patients increased 1. The ratio nonsyndromic versus syndromic remained almost similar. In approximately 2 of every 15 children the craniosynostosis is of the syndromic type, in 1 in every 8 of them other than Apert, Crouzon or Saethre-Chotzen syndrome.

The article does not make clear how syndromic and nonsyndromic craniosynostosis were distinguished. The associated anomalies in the syndromic types of craniosynostosis notably were anomalies of the face and of the extremities.

The London Dysmorphology Database version 1. More than half of the syndromes follow a Mendelian inheritance pattern; some are the result of teratogens including vitamin A and valproate. Chromosomal disorders are seen in a proportion of patients with a craniosynostosis including 9p-, 11q-.

Frias and Carey reviewed 4 population studies that established relations between the occurrence of minor anomalies and the risk of a major anomaly.

In the different studies, having 3 or more minor anomalies was associated with a From the same center in Paris, Lajeunie in published a series of patients with a nonsyndromic scaphocephaly of a total of craniosynostosis-patients admitted between and Also the remaining literature does not provide information on the ratio between nonsyndromic versus syndromic scaphocephaly.

Lajeunie 33 reports that most patients of scaphocephaly and trigonocephaly are nonsyndromic. A striking finding was 4. In , Kan 34 described that in none of the 13 patients with scaphocephaly an FGFR2 mutation was found in a comprehensive screening of the whole gene. Butzelaar 35 describes a pilot study in 30 consecutive patients with scaphocephaly, which retrospectively analyzed how many patients had consulted the Clinical Genetics department, what genetic tests were used, and what the test results were.

In addition, the parents were sent a questionnaire on risk factors. Maternal alcohol use and smoking habit did not differ from those in the general population. Prematurity was more often seen in the study group than in the general population.

None of the 30 included patients had craniofacial dysmorphias. In 4 of the 30 patients another anomaly was diagnosed nevoid basal cell carcinoma syndrome, defects of the cardiovascular system and of the urinary tract. Zeiger 36 investigated genetic and environmental factors associated with a higher risk of scaphocephaly in a group of 42 children from craniofacial clinics in the Baltimore-Washington area. Pathogenic mutations were not found.

The same mutation occurred in the patient's nonafflicted father. The significance of this mutation is not clear. Of a series of patients admitted between and in Paris, Lajeunie published an analysis of patients with a trigonocephaly. Fifty-three Thirteen of these 53 had a known syndrome; the diagnosis in the other 40 patients was unknown.

Also in the group of trigonocephaly patients the man:woman ratio was skewed, that is, 3. The proportion of patients with a positive family history was 5.

The proportion of twins in this series was 3 times higher than in the general population, and was higher than in the group of patients with a scaphocephaly 6.

Azimi 40 investigated 25 patients with trigonocephaly diagnosed between and In 16 patients it was an isolated anomaly, 2 patients in addition had a craniosynostosis of the sagittal suture without associated anomalies. Regarding the other 5 patients, in 1 the diagnosis of Say-Meyer trigonocephaly was made, in 11-cell disease and in another one Opitz-C syndrome. A diagnosis could not be made in 2 patients.

In 1 of 9 patients with a seemingly nonsyndromic trigonocephaly a mutation in the FGFR2 gene was found. Kress 42 found a FGFR1 mutation in 1 of 10 nonsyndromic trigonocephaly patients, who had or did not develop other dysmorphias. In , Kan 34 reported that in none of 17 patients with trigonocephaly a FGFR2 mutation was found in a comprehensive screening of the whole gene. The patients were classified into groups, a group of 40 patients with an isolated trigonocephaly and a group of 36 patients with associated abnormalities.

In 7 of the 36 syndromic trigonocephaly patients a 9p- or 11q-deletion was found. Four of these deletions were not detectable with conventional cytogenetic analysis. In a case-report, Van der Meulen 44 describes a trigonocephaly in a patient with Muenke syndrome.

In these types of craniosynostosis the coronal suture is involved; unilaterally in plagiocephaly, bilaterally in brachycephaly and oxycephaly. In oxycephaly there is also a craniosynostosis of the sagittal suture. Mulliken 45 performed molecular diagnostics in patients with a bilateral coronal suture synostosis, of whom 38 had been diagnosed with Apert, Crouzon, or Pfeiffer syndrome and 19 had no specific diagnosis.

In , Mulliken 46 reported results of molecular genetic diagnostics in 47 patients with a unilateral coronal suture synostosis. This was a prospective study in children admitted between and Theman:woman ratio was One patient was clinically diagnosed with craniofrontonasal dysplasia. Also the parents of the children were examined. Of the 47 children, 13 showed hypertelorism or had a parent with characteristics of craniosynostosis. All above anomalies were found in these 13 children. The known craniosynostosis syndromes, Apert syndrome, Crouzon syndrome, Pfeiffer syndrome, Muenke syndrome, Saethre-Chotzen syndrome, Carpenter syndrome, and craniofrontonasal dysplasia CFND, are characterized by craniosynostosis of one or both coronal sutures next to other birth defects.

In addition, mutations were found in 7 new exons. In patients with a syndromic craniosynostosis, genetic diagnostics is performed on the guidance of the syndrome diagnosis. FGFR1 mutations were not identified. Morris-Kay 53 reported a prospective study in patients born between and
Correction of craniosynostosis in childhood can cause relatively much blood loss. The risk of blood loss is higher in older patients and in corrective surgery of syndromic craniosynostosis. Next to the surgical and anesthesiologic challenges, we should take into account that syndromic patients may be associated with comorbidity. This is why optimal organizational conditions should be in place, before, during, and after the intervention.

This chapter deals with the specific risks involved in correction of nonsyndromic and syndromic craniosynostosis and recommendations are given to perform surgery as safely as possible. All members of the multidisciplinary team should be aware of possible comorbidity, that is, mostly in case of a syndromic craniosynostosis with compromised airway with or without OSAS.

Invasive monitoring is recommended in open procedures with expected severe blood loss. Invasive monitoring involves the use of a central venous line and an arterial line next to conventional monitoring capnography, ECG, pulse oximeter, FIO2, temperature, and urine output and precordial Doppler.

The major goals of invasive monitoring are timely recognition of serious problems that may occur in open craniosynostosis surgery, such as hypovolemia, hypotension, and electrolyte disturbances, as well as being able to check effectiveness of treatment of these problems. Venous cerebral congestion by hyperflexion of rotation of the head must be avoided. Usually, the patient is placed in a moderate anti-Trendelenburg position to reduce blood loss cave air embolism, see below under Complications.

Notably, in children with exorbitism the eyes should be well protected with eye cream, and pressure on the eyes must be avoided especially in prone position. Single administration of an antibiotic, after the induction, before start of surgery is standard procedure. Most recommendations are aimed at a balanced technique that provides for cardiovascular stability with the use of opioids and volatile agents next to relaxants.

The use of remifentanil infusion 0. An additional problem is that measurement of the lost volume intraoperatively is impeded by surgical technique and type of patient.

Few studies have been performed to optimize blood loss measurement and findings are mostly not conclusive. Infiltration of the skin with vasoconstrictors before the incision is much used but its effectiveness is disputed because of the greater degree of bleeding with treatment of the periostium and bone.
Controlled hypotension is rarely used because it has no clear benefits and can be disadvantageous in case of ICP and also in the anti-Trendelenburg position. The acute normovolemic hemodilution ANH technique is used before the surgical intervention after the child is brought under anesthesia. It involves removal of whole blood via the arterial or central line which is replaced with colloids or with crystalloids according to precise calculations and formulas. According to a B study, this technique in itself is not sufficient to reduce or avoid homologous blood transfusion in this type of surgery.

Duncan 82 found no difference in allogeneic transfusion rate when a cell saver was used and when not. Intraoperative antifibrinolytics. Aprotinin is no longer used after serious complications in cardiac surgery. Tranexamic acid: its use in itself is controversial, but is has been applied in combination with other methods.

Guidelines for the application of blood products are in place in every hospital as established by the local transfusion committees, and in conformity with the CBO Guideline Blood transfusion Venous air embolism VAE and the subsequent cardiovascular collapse can be prevented by a precise technique and by rapid application of adequate monitoring precordial Doppler, capnography, echocardiography, transcutaneous O₂—CO₂ monitoring, esophageal stethoscope, and venous central line.

Although Faberowski 86 reports an incidence of Meyer 88 reports a 2. The hemodynamic consequences of VAE are generally insubstantial, provided the patients are well monitored and measures are taken upon signs of VAE. As a result a massive blood loss the patient may develop relevant consumption coagulopathy and dilutional coagulopathy characterized, in principle, by depletion of soluble clotting factors. The situation changes when these children undergo a distraction osteotomy procedure and need to be intubated acutely in case of respiratory insufficiency, or when the distraction materials are being removed.

Evidence from the C level studies is conflicting on this issue. One of the 2 studies recommends immediate fiberoptic intubation, whereas the other study claims that the distractors exert minimal effect in the anesthesiologic conditions when certain factors must be taken into account the right screwdrivers and cutting pliers must always be available and that removal of the vertical bar allows for direct laryngoscopy.

The possibility of cerebral salt wasting syndrome should be considered when a patient develops hyponatremia after craniosynostosis surgery. Criteria for extubation at the end of the procedure, before transport to the ICU, are the following: rapid recovery of spontaneous and stable breathing, hemodynamic stability, normothermia, short to medium operation time with relatively little blood loss, and no continuous large blood loss through the surgical drains.

In principle all studies recommend ICU admission after open craniosynostosis surgery to continue volume management there and, if necessary, start artificial respiration.

Most studies on postoperative transfusion management report a tendency to overttransfusion. There is scarce literature on postoperative pain treatment after craniosynostosis surgery. In Rotterdam 2 double blind, placebo-controlled studies have investigated pain treatment in the first 24 hours after cranial correction for craniosynostosis. Van der Marel 95 treated 20 children with oral paracetamol and 20 with rectal paracetamol. The pain scores in the oral group were higher, but this effect disappeared with exclusion of the patients who had vomited after oral administration.

Despite the fact that Rectal administration of paracetamol was recommended. Prins 96 describes 12 children who received intravenous paracetamol and 14 children who received rectal paracetamol. This is indicative of more discomfort, possibly caused by more pain, and intravenous administration was recommended.

In the European literature, the use of paracetamol and NSAIDs is recommended if the blood loss is not significant with or without codeine phosphate British literature.

All textbooks emphasize, and this also appears from our own practice, is that the pain scores are surprisingly low. In patients operated on for nonsyndromic craniosynostosis, invasive monitoring can usually be discontinued the following day. Despite the proven effectiveness of erythropoietin and blood drawing for autologous transfusion, the use of these strategies is discouraged as they involve high costs and require repeated venipuncture, which is not child friendly.

Introduction of less invasive interventions is associated with less blood loss. As these, however, are very young children with a smaller circulating capacity, blood loss relatively is still significant. In selected cases admission to a medium care unit could be considered defined as a monitored bed providing for artificial respiration. The anesthesiologic goals in craniofacial surgery can only be realized in specialized pediatric centers, where multidisciplinary perioperative care is provided by a team composed of plastic surgeon, neurosurgeon, maxillofacial surgeon, pediatrician, pediatric anesthesiologist, pediatric intensivist, and specialized pediatric nurses with experience and means to manage and monitor this type of patients and where a sufficient number of children is operated on to keep the experience of the team at a high level.

Administration of EPO preceding the intervention, as well as collecting autologous blood for autotransfusion are advised against. Postoperatively a bed in a PICU must be available.

In less drastic interventions medium care may perhaps suffice, although 1 should be aware that these are mostly very young children with a smaller circulating capacity, in whom even slight postoperative blood loss must be monitored carefully. Invasive monitoring is recommended in the case of open procedures with expected severe bleeding. Overtransfusion in the postoperative phase should be prevented by adhering to the guideline on transfusion management.

Extubation at the end of the operation, before transport to the ICU, is possible in case of rapid recovery of spontaneous and stable breathing, hemodynamic stability, normothermia, short to medium operation time with relatively little blood loss, and no continuous large blood loss through the surgical drains.
The analgesic of choice for postoperative pain treatment is paracetamol. If this is not sufficient, an NSAID can be added even though this could increase the risk of postoperative bleeding. What are the indications for surgery in the different types of nonsyndromic craniosynostosis? What treatment is most indicated for the different types of nonsyndromic craniosynostosis? Nonsyndromic craniosynostosis can present in varying severity of the deformity of the skull.

Surgical treatment seems indicated on the basis of the variability in both the severity of the morphologic abnormality and in the occurrence of increased ICP may occasionally result in less strict operative indication on the basis of these parameters.

Many different surgical techniques have been described for the treatment of nonsyndromic craniosynostosis but the functional results of these techniques have not always been shown convincingly. In addition, the timing of surgery is debated. For patients with a synostosis of multiple cranial sutures in whom a syndromic diagnosis cannot yet be made, the method of treatment of syndromic craniosynostosis is recommended.

This chapter will place a focus on craniosynostosis of the sagittal suture scaphocephaly, metopic suture trigonocephaly, unilateral coronal suture frontal plagiocephaly, and lambdoidal suture pachycephaly.

The indications for treatment of craniosynostosis are the risk of ICP and the morphologic abnormality of the skull and face. The chapter on cognition and behavior provides evidence of absence of a relation between cognitive functions in children with a nonsyndromic craniosynostosis and surgical treatment or not, severity of the deformity, or age at operation. The prevention or treatment of the associated neuropathology is therefore no indication for surgery. The morphology can vary from very mild to an evidently abnormal skull shape.

In sagittal suture synostosis, compensatory growth of the other cranial sutures results in increasing deformity in the course of time, so that, for example, the frontal bossing in scaphocephaly can intensify after birth. Barritt evaluated a series of 44 children with scaphocephaly, of whom 34 did not undergo surgery.

This was common policy in this clinic at the time for children presenting with this condition at an age older than 6 months.

In these 34 children, the deformity in any patient did not seem to improve in the first 10 years. The presentation of trigonocephaly also varies from very mild to severe. Its mildest form is characterized only by a bony ridge as a result of premature fusion of the metopic suture.

In follow-up, cosmetic worsening is only rarely seen. The other end of the spectrum is characterized by a wedge-shaped forehead with hypotelorism and supraorbital retrusion on the basis of decreased growth of the forehead widthwise. Frontal plagiocephaly also shows some variation in phenotype, notably in facial asymmetry. Unilateral synostosis of a lambdoidal suture causes increasing asymmetry of the back of the head and particularly also of the face.

Increased ICP does not seem to be prominent in this type of synostosis, although reliable research on this issue is lacking see chapter 8. Since the first operative intervention for craniosynostosis, many surgical techniques for the various types of craniosynostosis have been described. A broad distinction is made between osteoelastic techniques and remodeling techniques.

In the first group bone is removed, enabling the developing and expanding brain to change the shape of the skull, in part because the adverse impulse to the growth direction of the skull also is removed.

From this technique evolved the remodeling techniques, because it was suspected that one cannot only rely on the self-correcting capability of the skull and the brain. These techniques are aimed at directly achieving the desired skull shape by a kind of reconstruction. Randomized comparisons of the different surgical techniques have never been performed.

For the different types of nonsyndromic craniosynostosis we will provide a review of the literature regarding the different techniques in these 2 groups, but also pay attention to the new developments in the past 10 years in a separate section. Ingraham described a technique using bilateral parasagittal strip craniectomies, in which the fused suture was not resected.

To prevent the problem of premature reossification of the skull bones he applied a layer of plastic to the bone rims. Successful treatment required early intervention, however, preferably in the first 2 months of life.

The results of this technique were confirmed by other authors. Albright presented a sagittal strip craniectomy with biparietal wedge osteotomies. The same procedure was performed by Marucci in 89 patients, but they noticed that 7 of the 89 patients later developed a cosmetically unsatisfactory vertex bulge, which they interpreted as a sign of possibly raised ICP or a new synostosis.

Also Florisson reported that some patients with scaphocephaly again showed signs of raised ICP postoperatively. Arm presented a technique, in which the usual 2. They explicitly try to make use of gravital force by keeping the infant strictly in supine position during sleep for a period of 3 weeks. Using this technique an average improvement of the CI of 8. All in all, many of the described techniques represent only slight modifications of previously presented techniques and not real conceptual changes.

The sagittal suture itself is not resected, and the temporoparietal bone is bent outwards. In this technique, the anteroposterior diameter of the skull is dynamically shortened by attaching the bone strip in the middle of the skull, which is shortened, to the frontal bone.

Thus, it is in fact also a compression technique, which might be associated with an increase in ICP. Only a global development quotient, however, was tested in very young patients, without inclusion of a control group, which raises great doubt on the reliability of this finding. In older children or in children with a more pronounced dysmorphology notably frontal bossing, a more extensive calvarial reconstruction is needed, with resection and reconstruction of the frontobiparietal complex.
The authors concluded that in this population this more extensive procedure had no adverse effect on postoperative growth rate of the skull or on intracranial volume. Lauritzen reported in on the first operations in which distraction was used to correct craniosynostosis. This method is eminently suitable for scaphocephaly in view of the unidirectional growth delay widthwise. The results are not worse, but also not better than those of the current techniques and the small numbers preclude valid comparison.

Also regarding the newer surgical techniques, randomized or comparative studies have not been published. Also in view of the complex three-dimensional dysmorphology seen in metopic synostosis, treatment by suturectomy solely is considered insufficient. Posnick analyzed the growth process on the basis of CT data in 10 patients and concluded that correction of the hypotelorism had remained insufficient. Selber found that the use of interpositional bone grafts in a series of 68 metopic children led to a decrease in the development of temporal dents.

In a population of 92 trigonocephaly children in which no bone graft was used, Van der Meulen, however, observed an automatic growth correction of the hypotelorism on x-ray, on account of which the use of an interpositional bone graft was judged to be unnecessary. From a photographic evaluation of 45 patients, Hilling concluded that a satisfactory esthetic outcome on the long term was largely dependent on a good initial reconstruction. A distinction is made between unilateral and bilateral frontal advancement techniques.

Some surgeons prefer the unilateral advancement technique so as to be able use the unaffected side as a reference for the degree of advancement, whereas others maintain that adequate advancement with good symmetry is only possible if the entire frontal bone and both orbital margins have been dissected. Barone and Jimenez report since the late 80s on endoscopic strip craniectomy of the affected suture. The benefits mainly lie in low morbidity, short hospitalization, and little need of blood transfusions in comparison with the classic fronto-orbital advancement technique.

The cosmetic outcome is reported as good, but it not adequately quantified. For this reason, the technique was adapted to an endoscopic unilateral fronto-orbital advancement technique.

This was applied in 10 patients, who did not develop any complications, did not need blood transfusions, and could be discharged home after 2 days.

The cosmetic outcome was assessed to be similar to that of fronto-orbital advancement. The number of publications on lambdoid suture synostosis is very limited, probably because of the very low prevalence. Unilateral lambdoid suture synostosis causes only relatively little volumetric restriction. The morphologic changes consist of an asymmetry of the face and an asymmetric basal occipital region. Kim reports a study comparing distraction versus remodeling surgery.

Among the patients were 3 children with brachycephaly treated with the distraction method and 4 with remodeling. The distraction method led to satisfactory calvarial expansion and an esthetically pleasing outcome. Advantages of distraction are the significantly shorter operation time, less bleeding, shorter stay at the neonatal intensive care unit NICU, and better skull shape.

A disadvantage of the distraction method is that children must have reached the age of 6 months. The authors are of the opinion that these downsides do not outweigh the benefits of distraction. Most children with a craniosynostosis are operated on in the first year of life. A direct evaluation of surgical outcomes is possible, but a final evaluation is not possible until the child has fully grown at adult age. As a surgeon's career spans perhaps 25 year, and at most 30 years, the surgeon can only at an advance career stage assess the result of his or her own work.

There may be a tendency to ascribe a poor result to a wrongly performed surgical technique, without questioning the technique itself. Furthermore, knowledge about the natural course of the condition with and without surgical intervention is usually lacking. This shortcoming undoubtedly is co-responsible for the fact that we see regular shifts in type of intervention in the course of decades and that paradigms change.

One and the same research group may recommend a certain approach in the 1 decade and disassociate oneself from it in the next decade. Only few solid objective results with such a long follow-up are available, whereas comparative studies of different surgical techniques with or without randomization have not at all been performed. Moreover, notably the larger series yield mixed data, because all craniosynostoses, including the syndromic types, are pooled.

One way to assess the outcome of surgery is looking at the operative morbidity and mortality. Most series are too small, however, to yield significant data. Morbidity is reflected in the complications that may occur and length of hospitalization. Although the first is a relatively objective score, the second is certainly not. Length of hospitalization strongly depends on the treating physicians, but also on the healthcare system itself. To illustrate this, for similar procedures the average length of hospitalization in North America is significantly shorter than in Europe.

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lateral sides of the forehead often appear to revert to a certain degree and temporal depressions may be seen, which however seldom are cosmetically unsatisfactory to the degree that revision surgery is justified.

Hilling evaluated the result of bifronto-orbital advancement in trigonocephaly from photographs of the patients pre- and postoperatively and after follow-up by a panel of independent assessors with the aid of a scoring form.

They concluded that the cosmetic result generally was good to very good, but also that the severity of the deformity does not influence the eventual outcome. In addition, they established that the obtained result is stable during time and independent of the age at operation 6 to 15 months.

Temporal depressions are ultimately the most frequent abnormalities at follow-up and cosmetically usually only little unfavorable. Van der Meulen performed standard x-rays of the skull before and after operation and in the course of time and noted that although in fronto-orbital advancement, the hypotelorism had not been actively corrected, autocorrection still occurred during time.

Assessment of the result of the various techniques applied in frontal plagiocephaly is highly subjective. Objective outcome measures are lacking and assessment therefore mainly consists of an evaluation of the achieved facial symmetry. Two of those patients underwent corrective ophthalmological surgery for strabismus as a result of a malformed orbit, whereas a third patient had rejected the same proposed intervention.

The earlier mentioned panel assessment of photographs by Hilling in trigonocephaly, was also applied by the same researchers in 59 patients with fronto plagiocephaly.

Also in this deformity, the temporal depression is the most striking finding at follow-up. Direct evidence for this supposition, however, is lacking. For that matter, this research group also concludes that the frequency of temporal depressions decreases with increasing experience of the surgeon. Lo specifically assessed the orbital morphology both of bone and soft tissue after surgical intervention and evaluated this by means of CT-scans.

They noted that the orbital morphology had genuinely improved, but also that complete symmetry had not been obtained. Furthermore, outgrowth, however, was normalized. The timing of surgery differs worldwide and is dependent, among other things, on the surgical technique used. In patients with boat-shaped head, it was observed that the initially improved CI index had slightly decreased 1 year after correction, whereas in fronto-orbital advancements, the frontolateral skull will again slightly recede a part reversal to the primary deformity.

This in general does not cause cosmetic problems, the more so because some surgeons in anticipation will apply overcorrection.

Thompson proposes that nonsyndromic craniosynostosis ideally is corrected within the first year of life, with scaphocephaly as a possible exception, corrected with early modified strip craniectomy. The largest series is reported by Marchac, nearly patients with all types of craniosynostoses were operated on in Paris. Pyron, J. Published by St. Louis : C. Mosby Co. About this Item: St. In , Emory recognized him for 50 continuous years of service in dentistry. Published by Georg Thieme Verlag Mrz Neuware - Anatomy for Plastic Surgery of the Face, Head, and Neck details the complex regional anatomy of the face, head and neck, providing plastic surgery and otolaryngology residents with a solid anatomical knowledge base.

There are many danger zones involved in operating on the head and neck, and the detailed knowledge of anatomy that readers gain from this reference will help them avoid the surgical mishaps that often result in patient disfigurement.

Key Features: Complex regional anatomy of the head and neck detailed with drawings, intraoperative photos and radiologic images Online access to videos in which authors walk readers through the anatomy of the face, head and neck Covers the latest anatomical topics, including arterial supply of the facial skin and sensory nerves of the head and neck This excellent anatomical reference will be read cover to cover by young plastic surgeons and otolaryngologists, as well as residents in these specialties.

More experienced surgeons will refer to it whenever they need to learn about an unfamiliar area of the head and neck. Published by Stuttgart Thieme Thieme Jun Neuware - This new text provides quick and easy access to a broad range of procedures performed by ophthalmic facial plastic surgeons. Brimming with beautiful illustrations, this exceptional resource features a brief review of key anatomy, background and management techniques in each disorder, and easy-to-understand surgical diagrams.

It also demonstrates the expertise of contributing leading specialists, who provide you with solid footing in each surgical intervention. Unlike many other texts, Oculoplastic Surgery addresses the fundamental importance of anatomy in developing an operative strategy. Through crisp line drawings of the face, lacrimal system, and orbit, each chapter systematically outlines the relationship of physical structure to the procedure.

Plus, you'll find step-by-step surgical approaches for functional and cosmetic problems, along with the latest advances in lacrimal surgery, ptosis repair, blepharoplasty, and more! Key features: - Anatomy introducing every chapter, providing you with the tools to completely understand and map out an operative approach - A practical, systematic orientation moving from external to internal structures - Nearly crystal clear illustrations that depict anatomy and techniques - The expertise of prominent physicians who offer a wide scope of diagnostic and treatment insights that can be tailored to your own situation.

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