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Craniopagus: the Suriname–Amsterdam conjunction

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Abstract *Objects:* A case of a Suriname female occipito-parietal to occipito-parieto-temporal craniopagus twins is described. The girls were transferred to the VU University Medical Center (VUmc) in Amsterdam, the Netherlands, for further diagnostics and to analyze whether surgical separation was feasible and ethically justifiable. The multifactorial aspects of different treatment options are discussed. *Methods:* The twins underwent multiple investigations by a multidisciplinary team. Advanced imaging techniques with 3D-CT scan, MRI and MRA scans, image fusion techniques and, most importantly, cerebral angiography with balloon occlusion tests were performed. *Conclusions:* Because of a shared venous ring, with preferential drainage to the left child, and which endovascular balloon occlusion showed could not be separated, surgical separation of the twins with a fair chance of survival without additional neurological damage and with prospects of a good quality of life was regarded as impossible. In accordance with the parents' wishes, the twins were not separated and offered optimal integral conservative treatment.

Keywords Computer-assisted image processing · Cerebral angiography · Balloon occlusion · Medical ethics · Rehabilitation · Conjoined twins

Introduction

Craniopagus is a rare congenital abnormality with an incidence of 0.6 per million births. With the assimilation of more cases of craniopagus in the recent literature and reports of more or less satisfying outcomes in difficult cases, separation should seriously be considered in each case [2–4, 6, 7, 11, 12]. We present extensive details of a case of occipito-parieto-temporal craniopagus and discuss why we decided against operative separation.

Case report

Description

Female twins, weighing 4,870 g, were born following an uncomplicated term fourth gestation in Paramaribo, Suriname by Caesarean section. The mother went into labor in an isolated bush village. Twins were not expected. After 36 h of ruptured membranes, due to insufficient engagement and meconium-stained amniotic fluid, she was urgently transferred to a general hospital in Paramaribo. The conjoined twins showed an occipito-parietal to occipito-parieto-temporal craniopagus without any other obvious congenital abnormalities (Fig. 1), but with initial asphyxia with bradycardia and hypotonia. One twin (Fig. 1, right child) initially showed respiratory and feeding problems due to extreme cervical flexion and torsion and was treated by careful positioning and feeding by gastric tube.

Skull ultrasonography and CT scan on the first day showed individual brains with normal supratentorial ventricles but hypoplastic cerebella with posterior fossa cystic malformations. Abdominal ultrasonography and total body X-ray showed no abnormalities.

The twins proved to be vital, showing an uneventful course during the first month of life. The parents were offered the support of social and pastoral workers and seemed motivated to have their daughters separated. As medical facilities in Suriname are inadequate to deal with uncommon and elaborate medical cases, the girls were transferred to the Department of Pediatric Neurosurgery and Pediatric Neurology of the VU University Medical Center (VUmc) in Amsterdam, the Netherlands under the provision of a long-standing bilateral agreement.

A multidisciplinary team, meeting weekly, was coordinated to prepare for transfer, diagnostic analysis, and coordination of all multifactorial aspects of integral care for a conjoined twin (see Table 1). In late September 2001, at 6 months of age, the girls, accompanied by both parents, were admitted to the VUmc.

Fig. 1 Craniopagus twins at 6 months of age. Note the extreme flexion of the neck in the right girl

Investigations

Diagnostic tests started after 5 days' isolation according to our local multiresistant staphylococcus aureus (MRSA) protocol. Weeks before admission logistics were extensively tested and adjusted at all locations using dummies. The girls were color coded with yellow (left child) and red (right child). All equipment, and even anesthesia personnel for each child, was coded accordingly.

Table 1 Multidisciplinary team and tasks. The team comprised 80 staff and personnel. *DMSA* dimercaptosuccinic acid scan, *EEG* electroencephalogram, *VEP* visual evoked potentials, *SSEP* somatosensory evoked potentials, *VUmc* VU University Medical Center

Discipline	Task—investigation
Neurosurgery	Coordination team for neurosurgical aspects, contacting media
Anesthesiology	Anesthesia for imaging studies
Neuroradiology	Imaging (X-rays, 3D-CT, MRI), (interventional) angiography, balloon occlusion tests
Nuclear medicine	Tracer studies (DMSA scan)
Physics and medical technology	Coordination of technical supplies, adapting hospital logistics. Image fusion in cooperation with Brain LAB and Dutch Cancer Institute
Pediatric neurology	Physical examination
Neurophysiology	EEG, VEP, SSEP
Pediatrics	Physical examination, pediatric daily care. Laboratory investigations
Pediatric nephrology	Renal function tests, blood pressure
Pediatric cardiology	Echocardiography and electro cardiography
Pediatric intensive care	Recovery after anesthesia, emergency care
ENT/laryngologist	Physical examination, intubation of right twin
Plastic surgery	Aspects of reconstruction (tissue expanders, etc.)
Cranio-facial surgery	Aspects of cranio-facial reconstruction
Laboratory	Standby in case of emergency
Technical service	Preparing logistics at different sites, checking equipment
Staff nurse ward, staff nurse operating theater	Management of nursing personnel schedule
Social worker	Supporting family, contacts with social care department Amsterdam
Public relations	Publicity strategy of VUmc
Audio visual communication	Recordings during admission in VUmc
Medical ethics/philosophy	Ethical aspects
Director, Medical Affairs, VUmc	Supervision

Table 2 Physical examination and investigations of conjoined twins at the age of 6 months. *ENT* ear, nose, and throat

	Left child (yellow)	Right child (red)
Neurostatus	Temporo-parietal fixation to sibling. Circumference of fusion 48 cm	Occipital fixation to sibling
	Comfortable supine position, prohibiting rotations and restricting movements with right arm	Comfortable supine position, prohibiting rotations. Anteflexion and dextroversion of head
	Symmetric face	Asymmetric face, left facial hypoplasia
	Inactive right corner of mouth	Inactive right corner of mouth
	Movement pattern with variations, progressive contact, and explorative behavior	Movement pattern monotonous and stereotypical, lack of explorative behavior
	Developmental age 4 months	Developmental age <3 months
Neurophysiology		
EEG	Independent pattern of one cerebrum	Independent pattern of one cerebrum
VEP	No registration possible	No registration possible
SSEP	No reliable registration	No reliable registration
Pediatric status		
Weight combined	10.6 kg	
Height	62 cm	57 cm
Heart rate	100 min^{-1}	160 min^{-1}, tachycardia
Blood pressure	Hypertensive, positional (140/90 mmHg), maximum systolic pressure 200 mmHg	Normotensive (100/60 mmHg)
Cor pulmones, abdomen	Enlarged liver	Normal
Infection	Multiresistant Klebsiella pneumonia (anus)	
ENT	Dysmorphic right ear and jaw	Positional cervical torsion, 90° torsion of epiglottis, partial obstruction of upper respiratory tract, difficulties in swallowing
Nephrologic status		
Diuresis	4 ml/kg/h	0.3–0.4 ml/kg/h
Creatinine clearance	55 ml/min/1.73 m^2, 80% of total	25 ml/min/m^2, 20% of total
Proteïnuria	20 mg/m^2/h (50 mg/m^2/h at age 1 year)	20 mg/m^2/h (13 mg/m^2/h at age 1 year)
Phosphate	2.5 mmol/l (High)	2.5 mmol/l (High)
Fractional magnesium excretion	9%	4%
Isotope (DMSA) scan	60% tracer excretion by kidneys	40% tracer excretion by kidneys
Cardiologic status		
Cardiac ultrasound	Left ventricular hypertrophy	Normal
Electrocardiography	Normal	Normal
Ophthalmologic status		
Fundoscopy	Normal	Normal

Physical examination

The relevant results of physical examination and investigations in the awake children are summarized in Table 2.

Anesthesia

With all personnel volunteering, diagnostic procedures under general anesthesia were performed on a Sunday, outside of the daily routine, in order to maximize patient privacy. The two anesthesiology teams were made up of two pediatric anesthesiologists, two senior residents, and two nurse anesthetists. Complete color-coded sets of separate equipment and medication were available for the twins. At pre-assessment it was realized that endotracheal intubation would possibly be difficult, especially in the right child because of the acute flexion and rotation of her neck. Intravenous glycopyrrolate (10 μg/kg) was administered before induction to dry mucous membranes. Because of the risk of losing a patent airway, anesthesia was induced by spontaneous respiration with the inhalation of volatile sevoflurane to the left child first, while supplying oxygen through a mask to the sibling. To detect a cross-circulation effect, end-tidal sevoflurane concentration was measured in the right twin. However, as both babies became agitated, they had to be inducted simultaneously. After ascertaining the option of mask ventilation, muscle relaxation was administered (atracurium 0.5 mg/kg, fentanyl 3 μg/kg). With the left child positioned higher than the right, the left child was intubated with a nasal tube with direct laryngoscopic control. Repositioning followed, with the right child higher than her sibling. The right girl could not initially be intubated by direct or flexible fiber-optic laryngoscopy as the larynx could not be exposed due to a 90° rotation of the epiglottis. Only after introduction of a laryngeal mask, passage of the flexible scope through the mask was possible and visualization of the larynx and intubation was successful. Anesthesia was continued with intravenous propofol, fentanyl, esmeron, and dormicum. Two Dräger Cicero machines with pressure-controlled ventilation were used at most locations. During transportation two Jackson–Rees systems and in the MRI unit two MR-compatible Dräger Titus anesthesia machines with Babylog ventilators were used.

During the initial day of anesthesia the twins remained cardiovascularly stable and were returned intubated for recovery to the pediatric intensive care unit.

Imaging (under general anesthesia)

The following examinations were performed: an X-ray of both cervical spines, cranial CT scan with axial slides of 2-mm slice thickness with 3D reconstruction, cranial MR imaging including MR angiography reconstruction. In addition, a cerebral angiography of both siblings was performed, puncturing both right femoral arteries using 4-French catheters. The different digital imaging data were additionally processed and analyzed using image fusion and

visualization techniques (VV 3D-Advanced, Brain LAB, Heimstetten, Germany) and a special fusion method developed in-house based on Mutual Information and volume rendering tools based on Open GL [9]. The elaborate and time-consuming work-up of these data showed that imaging technology combines science and art (see Fig. 6). After evaluation of the imaging studies, a cerebral angiography was repeated after 4 months, with catheterization of the right femoral arteries in both of the girls and the left femoral vein in the left girl; and this was combined with endovascular intravenous balloon occlusion tests.

Results of imaging studies

CT scan. This showed a large bony defect at the site of fusion, synostosis of the right parietal, occipital, and temporal bones of the left child with the right parietal and occipital bones of the right child (Fig. 2). Deformation of the right orbital was present in the left child together with even more severe deformation of the petrous bone, which is in close contact with the right petrous bone of the right child.

MR scan. Both supratentorial compartments contained completely individual, separate cerebra with an incomplete dural structure at the fusion site (Fig. 3a). In a common posterior fossa, four separate cerebellar hemispheres are connected to individual brain stems with both fourth ventricles communicating into a common cisterna magna or CSF space (Fig. 3b).

MR angiography. The arterial circulation of both the children was individually developed and seemingly separated. The venous drainage was shared and formed a venous ring in which transverse sinuses of both children participate with preferential drainage through the left child (Fig. 4).

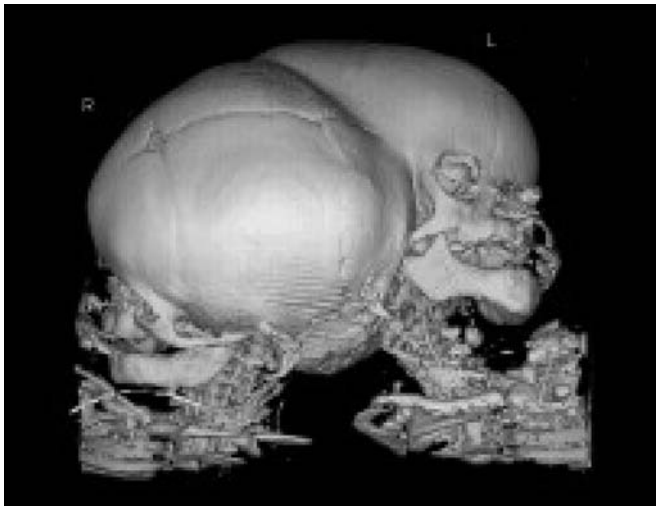

Fig. 2 Reconstructed image from the CT scan. A shaded surface 3D technique using a bone algorithm. The skulls are deformed and fused in both parieto-occipital regions. *R* right twin, *L* left twin

Cerebral angiography, arterial phase, of left child. The aorta is descending on the right side. Injection of the right common and internal carotid artery showed several branches from the proximal external carotid artery crossing the midline and connecting to the left external carotid artery of the right child (Fig. 5a). The internal and external carotid arteries of the right child are partially supplied by these collaterals. From the right ophthalmic artery a dural branch traverses cranially, crosses the

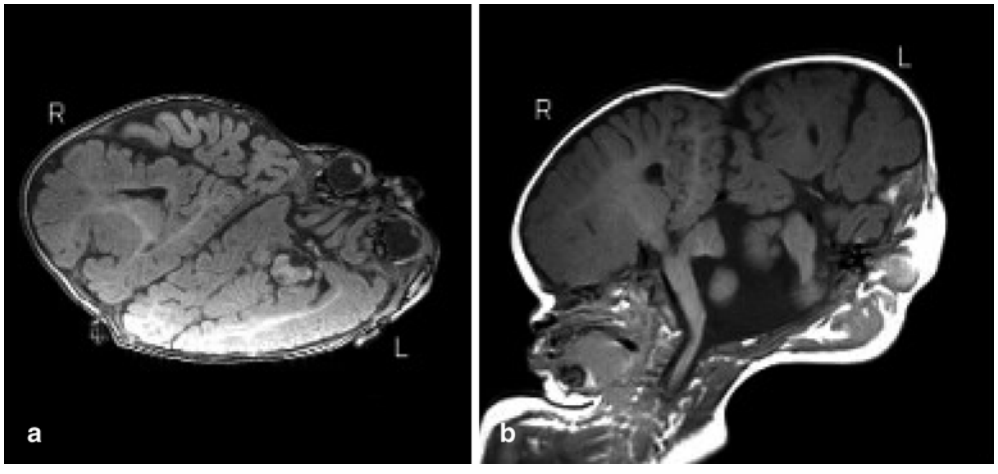

Fig. 3 **a** T1-weighted MR image in oblique plane for right child (*R*) and transverse plane for left child (*L*), showing two separate cerebra divided by an incomplete dura at the fusion site. Due to deformation of the skulls, the brains are slightly deformed. However, no developmental disorders are noticed. **b** T1-weighted MRI in sagittal plane for R and almost coronal plane for L, showing separate, individual cerebellar hemispheres and brain stems in a common posterior fossa

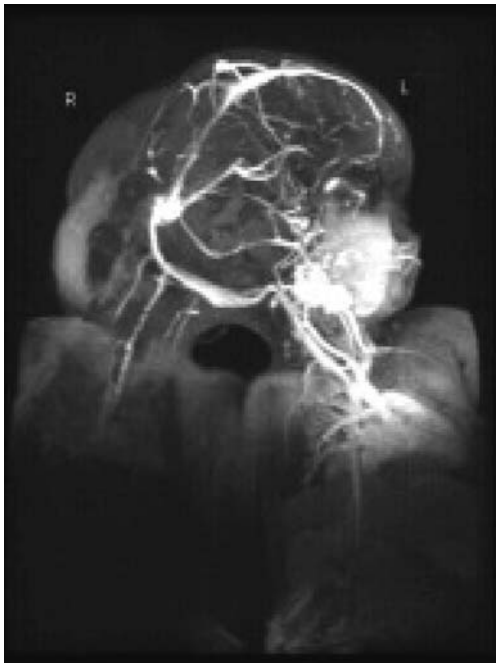

Fig. 4 Contrast-enhanced MR angiography, late acquisition during venous phase. The projection of the reconstructed image is similar to the angiographic images (Fig. 5c, d). Contrast was administered in the L. Although the complete cerebral venous system of L is visualized, drainage is through all four sinuses of both children, which make up a venous ring

midline to end caudally at the level of the left orbit of the right child, and supplies the left eye (Fig. 5b).

Cerebral angiography, arterial phase of right child. Injection of the left vertebral and left common carotid arteries. Normal supply to the posterior fossa and supratentorial left hemisphere. No cross filling.

Cerebral angiography, venous phase of both children. A venous ring is formed from the left and right transverse sinuses of the right child and the right-sided transverse sinus of the left child (Fig. 5c). The left transverse sinus of the left child is small and drains separately. The venous blood of the left child is shown to reflux into the sagittal sinus of the right child.

Cerebral angiography and balloon occlusion test. Introduction of a 4-French sheath into the right femoral arteries of both children and a 5-French sheath in the left femoral vein of the left girl. Introduction through the venous sheath of a microcatheter with a non-detachable balloon. The diameter of the inflated balloon was 8 mm. The balloon was positioned at three different sites in the shared venous ring to perform test occlusions (Fig. 5d). Contrast was injected separately into both children. No change in the venous drainage pattern was observed during the test occlusions. Both venous circulations remained connected. Even other venous collaterals previously not depicted were then visualized. It was concluded that the venous connections are too extensive to allow separation of the venous ring at one or two locations.

Advanced image processing techniques. The advanced 3D display and manipulating tools of fused sets of CT, MR T1-weighted and MR angiographic data facilitated further exploration of the unusual and abnormal cerebral blood vessels (Fig. 6a) beyond standard methods. The craniopagus case triggered the development of a new and useful registration (fusion) method to combine both the 2D X-ray angiographic data with the 3D MR angiographic and CT data (Fig. 6b).

Evaluation of results of investigations. After weekly multidisciplinary conferences and consulting different international colleagues there was general agreement that for surgical separation at least one child, most probably the right, would almost certainly have to be sacrificed, and there would be a very high risk of severe neurological damage or even death for the left child. For these reasons, and because the parents did not want to lose either of their children, it was decided not to separate. In addition, during the long period of clinical observation, both girls displayed a remarkable developmental evolution.

Discussion

Few clinical problems demonstrate the importance of an integral and multidisciplinary approach as when treating craniopagus. Dealing with the medical, (challenging) technical, social, and psychological aspects [1], as well as the medical ethical dilemmas, combined possibly with additional publicity, offers us the possibility of considering these individual, unique cases from different perspectives. After extensive analysis, a final decision whether or not to separate conjoined twins must be taken. Relevant aspects will be discussed.

Publicity

The birth of conjoined twins usually arouses the interest of the lay press, which may add to the difficulties of those involved in the care of these patients and this publicity may be harmful and even threatening to the privacy of the children, their parents and family, and at times even the hospital staff involved. In our case, the concurrent Twin-Tower disaster in New York, the parents' wish to respect privacy, and the media strategy of the VUmc, restricted media attention for our conjoined twins after their transfer to VUmc in Amsterdam to no more than a respectful 10-min documentary on national television and a short report in a newspaper.

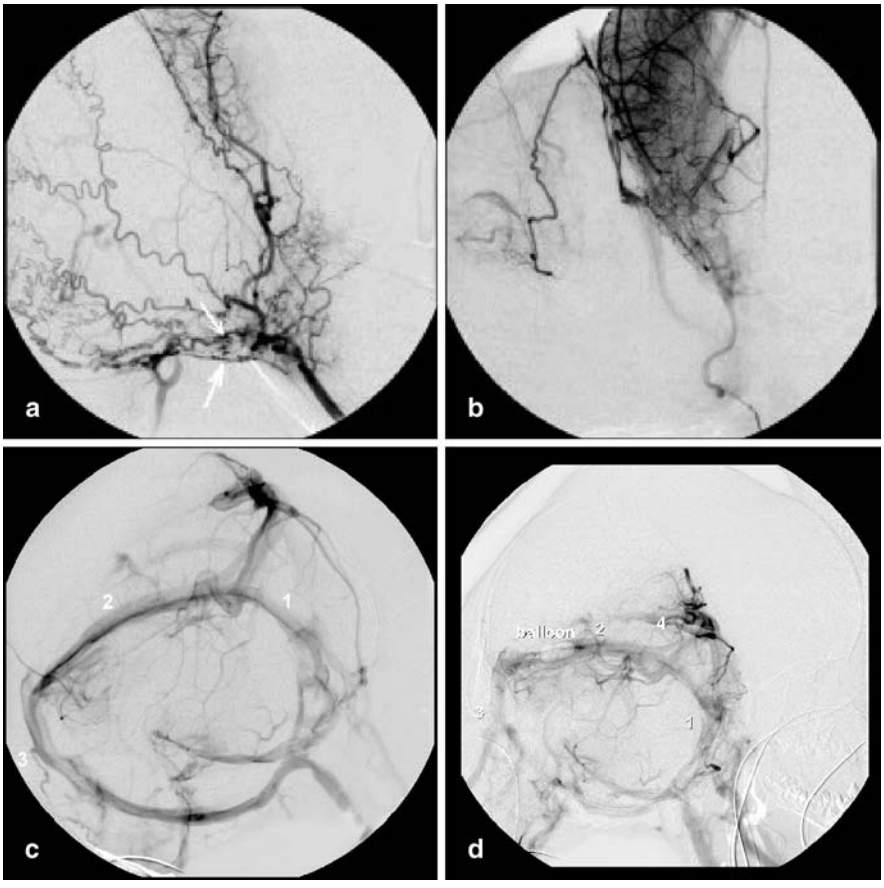

Fig. 5 **a** Digital subtraction angiography (DSA) with contrast injection in the right common carotid artery of L. Opacification of an extensive collateral network from the facial artery (*small arrow*) and the superior thyroid artery. These collaterals fill the territory of the left-sided external carotid artery of R and from here the common and internal carotid arteries (*large arrow*) are opacified. **b** DSA with contrast injection in the right internal carotid artery of L. Opacification of a dural branch from the ophthalmic artery crosses the midline and finally fills the left ophthalmic artery, depicting the contour of the left eye of R. **c** Venous phase after contrast injection of the left vertebral artery of R. A complete venous ring is seen. The left transverse sinus (*1*) and the right transverse sinus (*2*) of R and the right transverse sinus (*3*) of L form the complete shared venous ring. **d** Balloon occlusion test. Inflation of an 8-mm occlusion balloon in the right transverse sinus of the right child (*2*) and contrast injection through the left internal carotid artery. Despite the occlusion, the venous system, including the superior sagittal sinus, of the right child (*4*) is opacified by retrograde flow through different collateral channels

Socio-cultural aspects

The non-married parents lived in a bush village in Suriname and had three other healthy children. They were of low socio-economic and educational status, speaking only Creole and little Dutch. The birth of the conjoined twins was regarded as an ominous sign by the superstitious community they lived in. As a consequence, the family was no longer accepted in their village, and the father subsequently lost his job and had to leave to Paramaribo to find employment. A treaty exists between Suriname, a former Dutch colony and still a largely underdeveloped nation, and the Netherlands for treatment of uncommon elaborate or complex cases. Before transfer to Amsterdam it was discussed with the parents and medical colleagues in Suriname that, regardless of the outcome of the treatment in the VUmc, the family would have to return to Suriname and that arrangements for long-term rehabilitation should be prepared. As the parents would not be able to care for a conjoined twin or for that matter for two separate children with, possibly, double handicaps, the State of Suriname was responsible for and assured future rehabilitation, care, and education of the children *before* transfer, as Suriname lacks rehabilitation facilities, especially for children with double handicaps. However, the quality of future care in Suriname remained questionable and distrusted by the parents. During the hospitalization of the twins, relatives living in Amsterdam and the large local Suriname community provided an adequate social network. After the decision not to separate the twins, the family eventually received a permanent permit to stay in

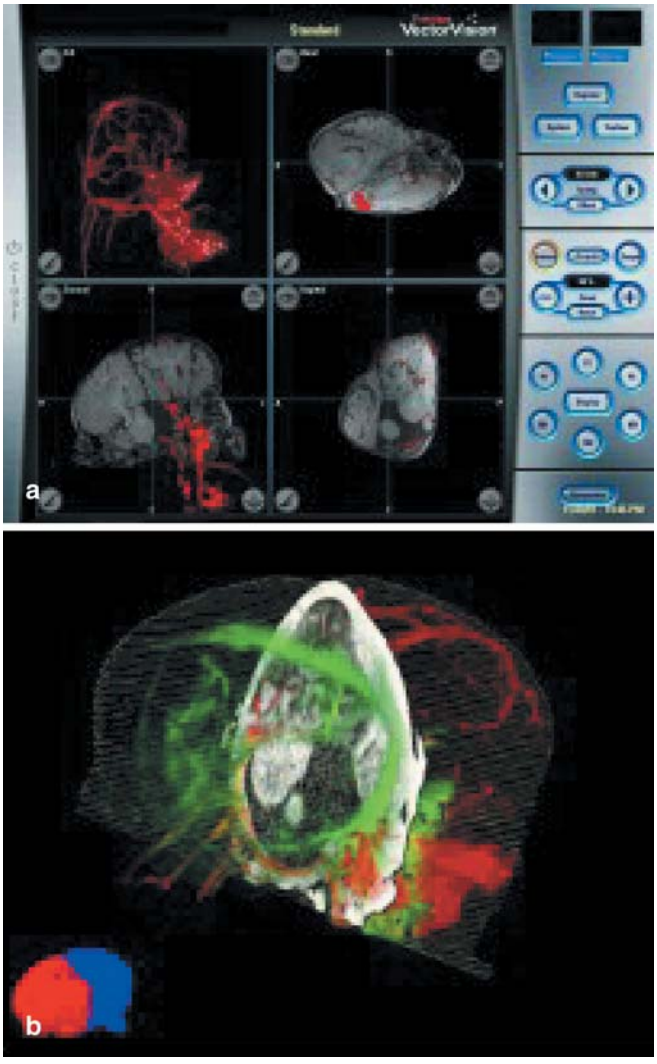

Fig. 6 a,b Advanced image analysis and visualization techniques. **a** 3D-rendered MR angiographic contrast-enhanced venous branches (*red*) can be visualized and manipulated within the anatomical MR-based or CT-based brain structure (BrainLAB, Germany). **b** Combined rendering of right and left segmented MR angiographic venous blood vessels and a CT slice of the separation plane between the two brains (Dutch Cancer Institute, Amsterdam)

the Netherlands, thereby assuring optimal future rehabilitation, educational facilities, and social care.

Ethical aspects

It is generally agreed that separation of craniopagus twins should at least always be considered [4, 10]. Existence in a conjoined state is not normal (from the point of view of *separate* individuals). In even the most understanding of social environments, craniopagus twins will experience severe embarrassment, ridicule, and disappointment, contributing to feelings of contempt and despair.

Ethical issues related to Suriname

In Suriname conjoined twins would be treated as social outcasts due to a superstitious belief that they bring bad luck. Thus, an eventual return to Suriname was an ethical issue in itself. As social acceptance and tolerance of children with deformities in Suriname is low in general and rehabilitation facilities are lacking, even the decision to start a trajectory of analysis with as a possible consequence surgical treatment, was only justified when future facilities for the twins and their family, regardless of outcome, were assured.

The choice between life and death

Moral theological considerations did not influence our process of decision making greatly. The mother was a protestant Christian and the father a so-called heathen. In Protestantism, life is measured by quality as well as longevity. In disjoining our twins, one child, the right, most certainly would have had to be sacrificed in order to give her sibling a chance to live. Protestant ethics regard such a separation as a positive act of saving and preservation of life and not as a negative act of destroying or sacrificing life, which is usually the Catholic position [10].

Issues related to quality of life

Even when theoretically separable, potential dilemmas arise if one twin should die because of the separation and the other survives, or when one or both children survive with severe neurological damage, or when both die. Regarding the prediction of survival and the quality of life of the one surviving sibling, another moral dilemma would arise: "What is *good* and what *good* can be done for the individual child?" If there had been a fair or even calculable chance of achieving a *good*, i.e., no (further) neurological damage and acceptable(?) quality of life for at least one child, we would have considered disjoining the twins. However, the neurological development of both the children proved quite favorable and the twins were comfortable. An adverse effect of the conjoined state on psychosocial development cannot be a decisive argument for separation, but should be weighed against the outcome of even the best technical separation. However, in our view, there was hardly any chance of a neurologically *good* outcome. The parents, who in time saw their daughters develop into lively toddlers, were no longer prepared to accept the loss or the occurrence of severe neurological damage to either of the girls. In the end, the children will have to cope with their fate of living an abnormal life in a conjoined state.

Issues related to illness or death of one twin

In case of life-threatening illness, the death of one of the twins would consequently mean the imminent death of the other. The deathbed of the longest surviving person can be very dramatic [5]. If this happens, especially when a twin is mature, moral, ethical, and legal aspects of euthanasia should have been discussed and cleared. An emergency operation to save one twin can be considered but is fraught with hazards. Another dilemma arises if, due to severe hypertension, one child, most likely the left twin, were to suffer severe end organ damage or a cerebro-vascular accident with intra-cerebral hemorrhage. Subsequent possibly severe neurological damage would be dramatic for the other, healthy twin. Surgical separation with the sacrifice of the neurologically damaged child should seriously be considered in this case.

Pediatric and nephrological aspects

The left and larger child has severe hypertension (maximum systolic blood pressure of 200 mmHg), which proved progressive during follow-up. On admission at 6 months of age, there already were signs of end organ damage in kidneys (proteinuria, impairment of tubular functions, calcium and magnesium wasting) and the heart (left ventricular hypertrophy). There were no signs of hypertensive retinopathy. The high blood pressure may be due to a lower venous resistance facilitating venous shunt to this baby with plethora and some fluid overload. This could explain the hyperfiltration seen in this girl. Intra-arterial blood pressure measurement in the ICU (after the imaging procedures under general anesthesia) showed a continuous difference in blood pressure between the twins. The difference in blood pressure could only be diminished when the left and larger baby was positioned higher than her sibling, thereby probably reducing relative fluid overload. As antihypertensive therapy of the left child was not thought to be possible without simultaneous effects on the sibling, positioning was the only therapeutic measure taken. Renal ultrasound showed normal kidneys. The DMSA scan showed 60% of total nuclear activity in the left and larger child and 40% in her smaller sibling. Individual renal function proved to be symmetrically divided. At 1 year of age, signs of renal damage proved progressive. Diuresis for the left child was 600 ml/24 h, whereas the right girl showed oliguria with 48 ml/24 h. In the hypertensive left girl proteinuria increased up to 50 mg/m^2/h, with microalbuminuria of 175 μg/min; the smaller child had less pronounced protein excretion (13 mg/m^2/h) and microalbuminuria (29 μg/min). Urine sodium excretion in the left twin (60 mmol/l) was normal and considerably higher than in the right girl (12 mmol/l), indicating pathological sodium retention with oliguria in the right girl. Progressive end organ damage may be reversible after separation, whereas life expectancy for the twins is expected to be reduced in the conjoined state [8]. During follow-up of the children at 3–4-monthly intervals as outpatients, their development progressed well. At the last follow-up, at 2.5 years of age, hypertension, especially in the left child, remains a problem, with blood pressures between systolic 178–228 mmHg and diastolic 98–156 mmHg and for the right child systolic blood pressures of 70–90 mmHg and diastolic pressures of 58–75 mmHg (Dynamap arm cuff measurement simultaneously in the sitting position, while asleep). With these blood pressures there is a considerable risk of progressive end organ damage. Anti-hypertensive treatment for the left child bears the risk of even more pronounced oliguria or anuria of the right child.

(Neuro)surgical aspects

On the basis of external appearance it is not possible to predict the exact extent of the sharing of anatomical structures. Our diagnostic work-up, using the most advanced imaging techniques, provided ample information to reliably weigh surgical decisions. The structures shared by our craniopagus were scalp and dura, but more importantly several arterial connections and significant common venous sinuses. As there were two individual brains our case was a type C, according to Winston's classification [12]. History has taught us that temporo-parietal and occipital junctions are related to the highest mortality and morbidity rates, but that outcome can also be related to the diameter of the joined area [3]. In our case both the area of conjunction (occipito-parietal to occipito-parieto-temporal), as well as the enormous circumference (50 cm) inferred a somber prognosis. Surgeons performing separations of craniopagi in the past were confronted with many hazards. Large scalp, skull, and dura defects predispose to wound problems because of insufficient tissue covering, CSF leaks, and infection. Air embolism during manipulation of venous sinuses, congestive and/or ischemic brain swelling and hydrocephalus have been encountered. Most precarious, however, is uncontrollable bleeding, usually from shared venous structures (and subsequently ischemia or venous infarction) [3, 7, 11]. The possible advantage of cardiopulmonary bypass and hypothermic arrest to provide a bloodless operative field inherently carries the disadvantage of other unforeseeable risks that may dramatically affect outcome [4].

In the stepwise approach towards eventual operative separation of our twins, the plastic and maxillo-facial surgeons would be confronted with an expected tissue defect of scalp, skull, maxilla, and mandibula, with a circumference of 50 cm in both children. To gradually gain scalp tissue by implantation of subcutaneous tissue expanders over the compressible skulls of the babies

Fig. 7 Prototype of externally adjustable vessel clamp (Zeppelin, Germany) mounted on a tripod to be secured with screws into the skulls of the twins if a gradual venous occlusion is a preliminary treatment option in a procedure to prepare for surgical separation

would take about 3 months in a procedure fraught with infection. Staged operations would then be scheduled for further cranio-facial reconstructions. The covering of very large dural defects with artificial dura substitutes, fascia or periost to cover brain tissue and control CSF leaks is another enormous challenge. Although individual brain tissue seemed separated on MR imaging, separation is not always as straightforward as it might seem [3]. The most formidable challenge would be the management of shared venous structures. The posterior part of the sagittal sinus and the transverse sinus of both children were shared. The preferential venous drainage of the right twin through the left sibling was by far the most complicated. Theoretically, a gradual occlusion at the point where the venous structure of the right twin drains into the circulation of the left child might establish separate, individual circulations, thus possibly preventing venous infarctions. However, control of the expected extent of thrombosis would be lacking. An externally adjustable (e.g., Moynihan) clamp mounted on a tripod to be fixed with screws into the skulls of the twins had already been designed and manufactured by our technical department (Fig. 7). Before proceeding with such a daring but questionable operation, the endovascular balloon occlusion test taught us otherwise. Balloon occlusion of the shared sinus at different sites proved that whatever interconnecting venous occlusions had been performed, the venous drainage would not be separated.

As a first procedure for the right twin, a venous bypass from the shared ring to, for instance, her hypoplastic jugular vein combined with reversible endovascular balloon or external clamp occlusion of peripheral venous cross flow(s) to her sibling could be a theoretical solution. The realization of a patent venous bypass in the delicate venous structures of a baby in whom flow velocities are low would be a great risk for the right twin in particular. The biggest problem would still be the collateral venous structures that make complete separation of venous drainage impossible.

As a successful separation, keeping both the children alive and in a good neurological condition would probably have more unforeseeable risks than those described, and the fact that the parents, who grew very fond of both girls, were concerned about losing their daughters, it was decided not to perform the operation.

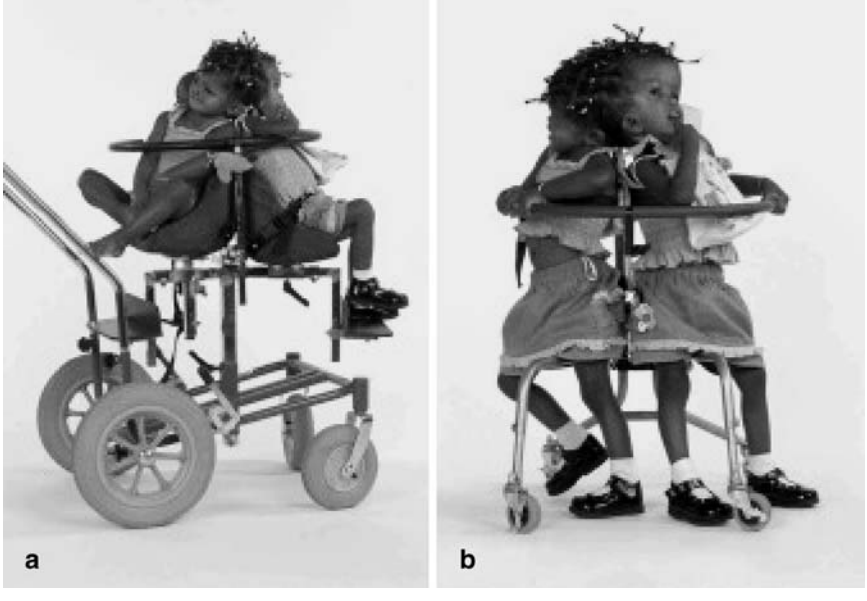

Fig. 8 **a** The twins at 2.5 years of age in a specially-designed push chair. **b** The twins in an ambulator enabling them to walk around

Rehabilitation aspects

As the family had in the mean time received a permanent permit to stay in the Netherlands, housing, social care, and home care were arranged. The children live at home with their mother, as the parents have separated. The twins follow a rehabilitation program in a day care center for 2 days a week with regular follow-up at the Department of Rehabilitation Medicine of the VUmc. The children are developing well neurologically with developmental milestones adjusted to the conjoined state. In the first year they learned to move around on the floor in a supine position and to roll over in an acrobatic way. To reach an upright position and to maintain balance in a sitting position was problematic, because of the craniopagus, and due to the relatively large, heavy heads and the extreme cervical flexion of the right child. However, they managed to stand up by pushing themselves up against a wall or pieces of furniture. At the age of 2.5 years they are walking a few steps with the hand-in-hand support of one person. It is to be expected that the twins will learn to ambulate without support. Fine motor skills are in accordance with age. Speech development of the right twin is delayed. The Department of Occupational Therapy of the VUmc was involved at an early stage in developing chairs and aids to mobility. First, adapted seats with removable tables and vertically as well as angularly adjustable head supports mounted on a circular wooden board on swivel wheels were constructed. Thus, the children were able to eat, drink, and play in a comfortable sitting position. A push-chair with adapted chairs and adjustable head support was made at a later stage (Fig. 8a). As the twins developed they were more and more eager to explore and wished to ambulate. An ambulator was constructed with vertically adjustable body-made seats and head supports mounted on a quadripod frame with four brake-loaded swivel wheels (Fig. 8b). Thus, the twins move in a crab-like fashion, using their legs to push the frame around, alternately taking the front position and having great fun. A circular rotator to help them walk more independently is under development. As the twins cannot see each other they will receive adjustable mirrors to communicate with each other.

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