CLINICAL OVERVIEW

Incidental sonographic diagnosis of neonatal carotid occlusion

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ABSTRACT

Cranial ultrasonography including colour Doppler can detect neonatal carotid flow problems at an early stage, even before symptoms occur. Different pathogeneses can be identified. The condition is more frequent than previously reported. If the circle of Willis is fully developed, this can prevent brain injury even in case of total carotid flow obstruction **Conclusion:** Screening of the carotid artery in critically ill neonates may detect complications of treatment at an early stage.

Imaging of the brain in preterm and term infants is important. It can be used for detection of abnormalities that may correlate with the clinical presentation or it can be performed to rule out or screen for abnormalities. In an optimal setting, magnetic resonance imaging (MRI) is the best neuroimaging technique due to its good spatial resolution, excellent soft-tissue contrast and quantification of brain injury (1–3). However, the use of MRI is hampered because it requires transport, sometimes sedation, such that critically ill neonates may not be stable enough to be scanned. Therefore, serial imaging is difficult.

Transfontanellar cranial ultrasound (CUS) is a safe and non-invasive method that can be performed bedside, even when a patient is unstable (4). CUS is nearly universally available on NICUs, which allows serial imaging. Therefore, CUS is the method of choice for brain imaging in daily practice in the neonatal intensive care unit. Extensive Doppler visualization of the circle of Willis is not performed routinely. In many cases, only Doppler measurements are performed in the superior sagittal sinus to check its patency and in the pericallosal artery to screen for luxury perfusion or increased resistance index. There is no additional training necessary to visualize the carotid arteries if the echographist is used to work with colour Doppler. One is able to screen for the carotid arteries in a coronal plane through the foramen of Monro.

Abbreviations

CUS, Cranial ultrasound; ECMO, Extracorporeal membrane oxygenation; ELBW, Extremely low birth weight; ICA, Internal carotid artery; MCA, Middle cerebral artery; MRI, Magnetic resonance imaging.

With six case histories, we illustrate that CUS with extensive colour Doppler of many intracranial vessels, including the circle of Willis, is feasible and that it can detect flow problems at an early, even presymptomatic stage.

In our centre, all infants are scanned according to standard protocol, including coronal and sagittal/parasagittal planes through the anterior fontanelle and colour Doppler visualization of different intracranial vessels (carotid arteries, basilar artery, Anterior cerebral artery, Middle cerebral artery (MCA), striatal arteries, both jugular veins and transverse sinus). We identified uncommon carotid pathology in six patients during 1 year (6 of 190 patients). In the context of this study, we will only focus on these six patients.

Images were obtained using the 8-MHz probe of an Esaote MyLab 70 (Genova, Italy). Ethical approval was obtained from the local ethics committee.

Case 1: A preterm infant born at 26 weeks of gestation was admitted in September 2010. He exhibited complications associated with infant respiratory distress syndrome, which was treated with mechanical ventilation. Initial routine CUS was normal. Because enteral feeding proved difficult, a Broviac catheter was inserted into the right internal jugular vein 11 days after birth, and CUS was performed 4 days after insertion. An unexpected phenomenon was observed with colour Doppler sonography (Fig. 1): the left internal carotid artery exhibited anterograde blood flow towards the circle of Willis, whereas the right internal carotid artery presented retrograde craniocervical flow. Axial images illustrated reversed flow in the A1 segment of the right anterior cerebral artery and



Figure 1 Composition of colour Doppler images (through the anterior fontanelle) on day 15 and day 44 after birth: the top row depicts coronal (in the middle) and sagittal scans of the flow direction in both carotid arteries, including velocity profiles. Normalization is illustrated in the bottom coronal image. Scanning is with the 8-MHz probe of an Esaote MyLab 70.



Figure 2 Axial colour Doppler image through the right sphenoid fontanelle on day 15. Adjacent schemes illustrate expected and abnormal flow direction.

forward flow through the right posterior communicating artery from the P1 segment of the posterior cerebral artery (Fig. 2). Ultrasound of the neck documented thrombus in the common carotid artery proximal to the bifurcation. Because the circle of Willis was fully developed and competent, flow was maintained and a stroke did not occur. The surgeon noted that the right common carotid artery had been accidentally punctured. MRI was performed and included 'time-of-flight' images on day 25. These data showed asymmetrical signal intensity in the carotid arteries. After team debate, we decided not to treat this thrombosis due to the risk of embolism. On day 20, however, a thrombosis in the left transverse sinus was documented and treatment with low molecular weight heparin (LMWH) was started. By week 7, the right internal carotid artery had regained forward flow (Fig. 1). Serial CUS showed no focal lesions until discharge at 35 weeks postmenstrual age.

Case 2: A preterm infant born at 27 weeks of gestation was admitted in April 2011. She needed ventilator support by IRDS, and intubation proved difficult. Three days of respiratory support was given non-invasively. On day 5, an urgent tracheotomy was necessary because of respiratory insufficiency, difficult intubation by laryngeal oedema and velopharyngeal hypotonia. The procedure was difficult, and the surgeon ligated a vessel in the operating area. On day 11 after birth, standard CUS revealed retrograde flow in the right internal carotid artery. Axial images were comparable with Fig. 1. In retrospect, the vessel ligated during the procedure must have been the right internal carotid artery. Genetic counselling and DNA diagnostics confirmed 22q11 deletion.

Case 3: A preterm infant born at 31 weeks of gestation was admitted in May 2011. Pregnancy and delivery were uncomplicated. CUS on admission documented extensive intraventricular haemorrhage complicated by venous infarction of the right anterior terminal vein. Serial CUS illustrated a transient absence of flow in the left carotid artery. Adjuvant images were obtained through the sphenoid fontanelle, and compensatory flow through the circle of Willis was not seen. A Rickham drain was inserted for posthaemorrhagic ventricular dilatation. A few days after insertion, the head circumference remained stable and further tapping of cerebrospinal fluid was not necessary.

Case 4: A near-term infant was admitted in June 2011 because of seizures within 24 h after birth. Pregnancy and delivery were uncomplicated. CUS showed extensive intraventricular haemorrhage in the right ventricle without venous infarction and a transient absence of flow in the left carotid artery. He was treated with anti-epileptic drugs and frequently monitored for posthaemorrhagic ventricular dilatation; only one lumbar puncture was indicated.

Case 5: A preterm infant, first of twins, born at 34 weeks of gestation was admitted in April 2011 because of respiratory insufficiency due to pneumothorax. The second postnatal day, he was transferred to the paediatric intensive care unit for extracorporeal membrane oxygenation (ECMO) to treat persistent pulmonary hypertension. CUS after ECMO failed to document brain injury, whereas retrograde filling of the internal carotid artery was present. His course was complicated by severe bronchopulmonary dysplasia.

Case 6: A near-term infant, second of twins, was admitted because of seizures within 24 h after birth. Pregnancy and delivery were uncomplicated. CUS on admittance did not reveal haemorrhage or other lesions. The MR-DTI sequence documented stroke in the posterior trunk of the left MCA and a perforator stroke in the left thalamus. Additional colour Doppler imaging after MRI showed absent flow in the left carotid artery, with compensatory flow to the left MCA along the anterior communicating artery (Fig. 3). Because of a suspected thrombus in the carotid artery, the patient was treated with heparin. After 5 days, the left carotid artery regained forward flow.

The history of the first two patients illustrates how acute complete obstruction of the common carotid artery can be missed in an extremely-low-birth-weight (ELBW) preterm infant due to a lack of symptoms because of sufficient

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Figure 3 Absent flow in the left carotid artery, with compensatory flow to the left Middle cerebral artery (MCA) along the anterior communicating artery.

collateral flow through the circle of Willis. These observations also demonstrate the potential of CUS to document changes in the condition of an ELBW infant. With transfontanellar colour and power Doppler imaging, we investigated flow in vessels high up in the neck and documented the competence of the anterior and posterior parts of the circle of Willis to detect presymptomatic flow problems. The images of patients 1, 2 and 5 suggest obstruction of the right common carotid artery and retrograde filling of the internal carotid artery from the circle of Willis. The explanation for these observations was that obstruction was located below the bifurcation, which conserved anterograde flow in the right external carotid artery. If an internal carotid artery is obstructed, the contralateral carotid artery may augment flow and perfuse the area supplied by the affected anterior cerebral artery; this mechanism, together with forward flow through the posterior communicating artery ipsilateral to the obstruction, sustains perfusion of the middle cerebral artery on the affected side (5, 6) (Figs. 1 and 2). The images of patient 6 were compatible with Internal carotid artery (ICA) thrombosis between the bifurcation and the circle of Willis.

Some discussion of the variability of the circle of Willis is warranted. Our findings suggest that colour Doppler and power angiography data allow to study the circle of Willis in appropriate clinical settings (e.g. before and after ECMO, jugular or carotid cannulation and aortic surgery). Thomas Willis described his 'circle' in 1664: it is generally a 9-sided polygon that is completely formed in the 52-day-old embryo (2). Variations result from different mechanisms (7, 8), which may be developmental (genetic or acquired), but may also result from structural changes acquired during adulthood (3). The definition of a normal circle is debatably based on a study by Alpers et al., who found a 52% incidence of normal circles in a postmortem population without brain injury (5). The circle can be incomplete because of a lack of polygonal components or can have more than nine vessels forming the polygon (9); vessels may also be present as expected but exhibit asymmetric calibre and flow. ICA agenesis or hypoplasia is most often unilateral and rare (< 1/10,000 individuals). Perfusion of the MCA on the affected side may occur through a

transcavernous collateral from the contralateral ICA, from intact parts of the circle of Willis or from anastomoses with the external carotid artery (10,11). The main vascular supply of the brain during bilateral agenesis of the ICA is the vertebrobasilar system (12–14). CUS is useful in detecting unilateral carotid artery agenesis (10,11,15,16).

The remarkable phenomenon of acute competence of the circle of Willis is also observed in many neonates who are treated with arteriovenous ECMO, where the right carotid artery is cannulated (patient 5). This procedure may predispose such patients to brain injury; the incidence of cerebrovascular injury ranges from 10 to 52% (17).

Patients 3, 4 and 6 had different flow patterns and did not show retrograde flow in the carotid artery. In these patients, flow was absent or seriously decreased in the left carotid artery, which spontaneously restored within days. Cerebral vasospasm as complication of subarachnoid haemorrhage is well known and can result in neurologic deficits. Only three paediatric cases are reported of intraventricular haemorrhage leading to vasospasm (18–20). We hypothesize that patients 3 and 4, both with intraventricular haemorrhage, had no visible Doppler signal of the left internal carotid artery due to (transient) vasospasm. Patient 6 had an ICA thrombosis proximal to the bifurcation.

Puncture of the carotid artery, seen in patient 1, is a complication of jugular vein catheter insertion, and the incidence is approximately 8.5–23% (21,22). However, (complete) obstruction of the common carotid artery has not yet been reported in a preterm infant, although Broviac catheters are frequently inserted.

Management of cerebral arterial thrombosis is still controversial. Recurrent arterial ischaemic stroke or carotid dissection is treated with LMWH at least for six weeks (23). However, treatment strategies for carotid thrombosis are not specifically described in these guidelines. Therefore, we decided not to treat in the acute phase because of the risk of embolism.

In conclusion, targeted CUS including colour Doppler can detect flow problems at an early, even presymptomatic stage. Different pathogeneses can be identified for carotid flow problems. The neonatal circle of Willis is competent to prevent brain injury if fully developed. Screening of the carotid artery in critically ill neonates can detect complications of treatment at an early stage. There is a place for routine insonation of the large neck vessels in critically ill neonates.

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