Perfusion MRI demonstrates crossed-cerebellar diaschisis in sickle cell disease

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Case report:
Perfusion MRI shows crossed-cerebellar diaschisis in sickle cell disease


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running title: PW-MRI shows diaschisis in SCD

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Abstract
Arterial spin labelling is a fully non-invasive magnetic resonance perfusion imaging method ideally suited to paediatric perfusion imaging. Here we present the case of an eight year old male patient with sickle cell disease and extensive right hemispheric cerebral infarction, with crossed-cerebellar diaschisis apparent on arterial spin labelling perfusion MRI. To our knowledge, this is the first case of crossed-cerebellar diaschisis demonstrated with arterial spin labelling, demonstrating the potential value of perfusion MRI in the clinical evaluation and follow-up of crossed-cerebellar diaschisis and the suitability of arterial spin labelling methods for routine perfusion imaging in paediatric patient groups.

Introduction
Arterial Spin Labelling (ASL) is a fully non-invasive MR perfusion imaging method which utilises magnetically labelled arterial water as a freely diffusible endogenous tracer, enabling perfusion to be quantified from 2 sets of images acquired with and without prior spin labelling. This method has been used in a number of clinical and research studies, [1-10] and has recently been implemented as part of a routine brain MR imaging protocol in some centres [11-13]. ASL perfusion MRI is particularly suitable for paediatric populations because it does not involve exposure to ionising radiation or injection of a contrast agent. In children, the higher perfusion rates also provide a strong ASL signal, while in certain adult patient populations the low signal to noise ratio relative to other perfusion imaging techniques represents one of the main limitations of spin labelling methods.

ASL perfusion imaging holds particular promise for the assessment and monitoring of paediatric stroke. In addition to regions of localised ischemic damage, stroke also produces areas of hypoperfusion in distant areas resulting from interruption of the afferent/efferent fibre pathways providing excitation to those regions. This phenomenon of diaschisis can be seen both contra- and ipsi-lateral to the area of primary stroke, depending on the morphology of the affected fibres and the age at the time of cerebral insult [14], but is most often noted in the contralateral cerebellar hemisphere resulting from alteration of excitatory or inhibitory inputs through the corticopontocerebellar fibres. While this crossed-cerebellar diaschisis (CCD) is
typically reported in the context of large hemispheric infarcts, it has also been reported to arise from small white matter strokes [15,16], migraine [17], encephalitis [18], tumours [19], and epilepsy [20]. Within the context of acute stroke, crossed-cerebellar diaschisis has been observed to correlate positively with the volume of primary hypoperfusion and inversely with stroke outcome, such that the presence of diaschisis in the acute stage appears to be associated with a less favourable outcome [21]. The present report describes a case of crossed-cerebellar diaschisis in a patient with sickle cell disease and right hemispheric infarction, which was revealed with arterial spin labelling MR perfusion imaging.

**Case report**

This report describes the case of an eight year old male with severe right hemispheric stroke and resulting left sided weakness secondary to cerebrovascular disease associated with sickle cell disease. Prior to stroke onset the patient was neurologically normal. Initial CT performed acutely on day 1 and day 3 at an outside institution (figure 1) revealed an extensive area of low density in the right cerebral hemisphere involving the right middle cerebral artery (MCA) territory. MRI performed at day 2 demonstrated an extensive area of cortical T2 hyperintensity involving the entirety of the right MCA territory in addition to T2 hyperintensity of the right basal ganglia. Further cortical T2 hyperintensity was seen in the left frontal lobe in parasagittal watershed regions and mature small infarcts were noted in the left lentiform nucleus. Although no angiographic sequences were performed, the intracranial portion of the right internal carotid artery appeared markedly attenuated in calibre on structural MRI.

The patient was treated neurosurgically in the acute phase by decompressive craniotomy and referred to the regional neurosciences centre for further imaging. A follow-up MRI performed after 4 ½ months demonstrated extensive established infarction involving the entirety of the right MCA territory including the corpus striatum, with additional mature infarction involving the right anterior and posterior cerebral artery territories and ischaemic change in parasagittal watershed regions and basal ganglia on the left side (figure 2). Regional volume loss with sulcal dilatation and ventricular prominence was noted. MR angiographic imaging demonstrated severe narrowing of the distal right internal carotid artery and a stenosis at the origin.
of the left MCA, with flow in the right anterior and middle cerebral arteries originating from the anterior communicating artery. Arterial spin labeled perfusion images were acquired with a 1.5T GE TwinSpeed HD.x MRI scanner (GE Medical Systems, Milwaukee, WI, USA) using a pseudo-continuous labelling scheme with a 3D interleaved spiral fast spin echo readout [22]. 64 axial slices were collected with a repetition time (TR) of 5.5 seconds and an echo time (TE) of 25 ms, a slice thickness of 3 mm, a field of view of 24 cm, and an acquisition matrix of 64x64. The ASL perfusion images revealed an extensive area of hypoperfusion corresponding to the area of primary infarction, with a corresponding decrease in perfusion in the left cerebellum consistent with crossed-cerebellar diaschisis (figure 3). There was minor left cerebellar hemispheric volume loss but the degree of perfusion abnormality appeared slightly out of keeping with the severity of volume loss.

Moderate recovery was noted on clinical follow up and the patient could walk independently with the aid of a foot orthosis, although there was persistent left hemiparesis, particularly distal to the wrist, and left homonymous hemianopia. Initially left handed, the patient now showed signs of right handedness. There were no obvious cerebellar signs. Cognitively there was some disinhibition but no other major concerns. The patient has since then had an uneventful EDAS (encephalo-dural-arterial-synangiosis) procedure on the left side and no subsequent strokes.

**Discussion**

Crossed-cerebellar diaschisis (CCD) is thought to arise from reductions in the afferent inputs to crossing corticopontocerebellar fibres, resulting in functional deactivation and decreased perfusion in the cerebellar hemisphere contralateral to the primary site of hypoperfusion. The severity of CCD appears to represent a possible marker for recovery and outcome, and has also been shown to be responsive to treatment [16,21].

To date the vast majority of CCD cases have been reported with Single Photon Emission CT (SPECT) and Positron Emission Tomography (PET), but one previous study examined the incidence of CCD in a series of stroke patients with Dynamic Susceptibility Contrast (DSC) perfusion MR imaging [23]. Like DSC-MRI, ASL is advantageous in that it does not involve exposure to ionising radiation, can be acquired with relatively high spatial resolution, and can be applied in conjunction
with structural, diffusion-weighted, and angiographic MR imaging, thereby enabling a more extensive evaluation of tissue morphology and function within a single scanning session. However, ASL also does not require injection of intravenous Gadolinium, making it fully non-invasive and ideally suited to paediatric populations and groups of patients with impaired renal function. ASL perfusion MRI may therefore represent a valuable potential tool for investigating the clinical and prognostic significance of crossed-cerebellar diaschisis and further elucidating the relationship between diaschisis and recovery.
References


Figure 1. Acute CT (day 3) and T2-weighted MRI (day 2), demonstrating a large acute right middle cerebral artery territory infarct with mass effect and midline shift. Note the area of old infarction within the left lentiform nucleus.

Figure 2. Follow up T2-weighted MRI and corresponding CASL images after 4 ½ months show expected evolution of the large established right MCA infarct with a corresponding perfusion deficit.
Figure 3. Follow up axial T2-weighted and corresponding CASL perfusion MRI images show a relatively normal cerebellum on structural imaging other than very subtle fissural prominence on the left, but a clearly obvious perfusion defect within the left cerebellar hemisphere, in keeping with the phenomenon of crossed cerebellar diaschisis.