

blindness, dysphagia, mental deterioration and milestone regression.

MRI has been shown to play a fundamental role in diagnosis and follow-up imaging of children with Krabbe's disease. Provenzale *et al.*² showed good correlation of MRI findings (using Loes scores³) with clinical scoring systems. Centrum ovale, basal ganglia, thalami, internal capsule, cerebellum and brainstem are all areas that demonstrate T2 and FLAIR sequence hyperintensity. Thalamic involvement is a useful sign to differentiate from metachromatic leukodystrophy.⁴ Optic nerve hypertrophy and/or enhancement, as well as enhancement of other cranial nerves, is a feature of the disease.⁵ Severe progressive atrophy, involvement of U-fibres, cortical blindness and seizures are usually

late phenomena. Diagnosis is made by detecting enzyme deficiency in leukocytes or cultured skin fibroblasts. Treatment includes bone marrow transplantation, supportive therapy and physiotherapy.

1. Cheon J, Kim IO, Hwang YS, et al. Leukodystrophy in children: A pictorial review of MR Imaging features. *RadioGraphics* 2002;22:461-476.
2. Provenzale JM. Correlation of neurodevelopmental features and MRI findings in infantile Krabbe's disease. *AJR* 2009;192:59-65.
3. Loes DJ, Hite S, Moser H, et al. Adrenoleukodystrophy: a scoring method for brain MR observations. *AJNR* 1994;15:1761-1766.
4. Candy S. An Approach to White and Grey Matter Disease in Children. In: *Baby Steps into Paediatric Neuroradiology*. Andronikou S, Wieselthaler N, Kader E (eds). 1st ed. Pretoria: SAMA Press, 2004.
5. Jones BV, Barron TF, Towfighi J. Optic nerve enlargement in Krabbe's disease. *AJNR* 1999;20:1228-1231.

Paediatric vascular imaging

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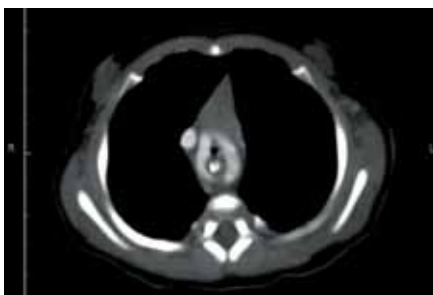


Fig. 1.

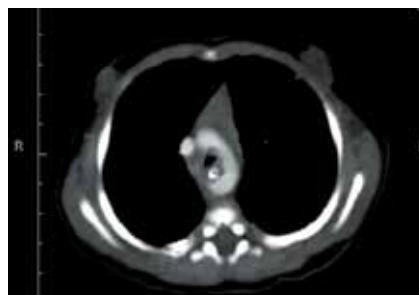


Fig. 2.



Fig. 3.



Fig. 4.



Fig. 5.



Fig. 6.

Presentation

An early neonate presented with stridor, and the following images were obtained. Figs 1 and 2 are axial post-contrast CT angiogram images at the level of the aortic arch. Note a nasogastric tube *in situ*. Figs 3 and 4 are volume-rendered 3D reconstructions of the CT angiogram from the level of the aortic root to the great vessels. Fig. 5 is a coronal reformatted image through the abdomen. Fig. 6 is a virtual bronchoscopy image of the tracheobronchial tree.

Describe the relevant findings and provide the most appropriate clinical diagnosis. Please submit your response by email to Dr Misser at misser@lakesmit.co.za not later than 1 February 2012. The winning respondent will receive a R1 000 award from the RSSA. A detailed diagnosis and discussion will be presented in the next SAJR.