

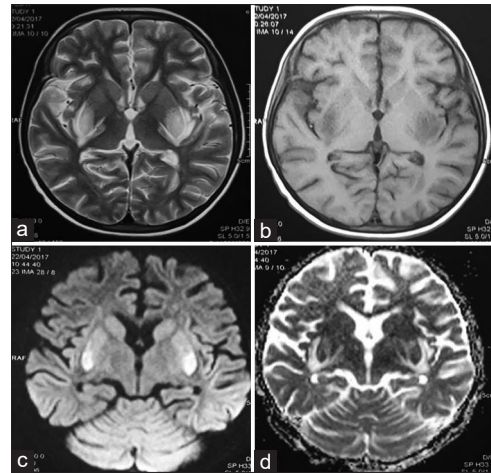
## Lentiform Fork Sign in a Child on Hemodialysis

Sir,

Lentiform fork sign is a recently described radiological finding attributed to oedema of basal ganglia.<sup>[1]</sup> This sign has been described in clinical situations with metabolic acidosis including poisonings and uremia.<sup>[2-5]</sup> and in a child with dialysis disequilibrium syndrome.<sup>[6]</sup> Delayed appearance of this MRI sign after several weeks of dialysis has not been reported so far.

A 9 and a half year old boy presented with end stage kidney disease. His native kidney disease was unknown and he was initiated on hemodialysis. After 12 dialysis sessions, he developed fever and chest signs. CT scan of the chest showed near total collapse consolidation of right middle and lower lobes and patchy consolidation in apical segment of left lower lobe. In spite of intravenous antibiotics, he became hypoxic and was started on supplemental oxygen. Echocardiography revealed severe left ventricular dysfunction and dobutamine infusion was started. After 27 sessions of hemodialysis, he developed altered sensorium with staring look, no verbalization and inability to obey commands. There was hypotonia and grade 1 power with absent deep tendon reflexes in all 4 limbs. Bilateral plantar reflex were mute. Meningeal signs were negative and pupils were equal and reacting to light. There was no respiratory muscle or cranial nerve involvement. Arterial blood gas analysis showed mild metabolic acidosis (PH – 7.37, PCO<sub>2</sub> – 26.7mm of Hg, PO<sub>2</sub> – 65 mm of Hg, HCO<sub>3</sub><sup>-</sup> 15.1meq/L, Base excess- -3.1meq/L). MRI of the brain showed unusual findings [Figure 1] with bilateral T2 and FLAIR hyperintensities of lentiform nuclei, with a brightly hyperintense rim surrounding both putamina that looked like a fork, known as lentiform fork sign. In spite of daily hemodialysis, his condition worsened with development of multiple episodes of focal seizures, dystonia of all four limbs, bilateral ankle clonus, and brisk deep tendon reflexes of upper and lower limbs and he was intubated in view of irregular breathing. He developed hypotensive episodes during hemodialysis sessions and hence, continuous ambulatory peritoneal dialysis (CAPD) was started. Over the next week he improved with the appearance of spontaneous movements of limbs and improvement in sensorium and verbalization and was extubated. Over the next weeks, he continued to improve with intensive physiotherapy and speech therapy. MRI brain done after 3 months was normal with resolution of lentiform fork sign.

Lentiform fork sign is attributed to vasogenic oedema of bilateral basal ganglia.<sup>[1]</sup> The components of the lentiform fork has been described by Kumar *et al.*: (1) the lateral arm, formed by the edematous external capsule and extending



**Figure 1: MRI Brain: (a) bilateral T2 and FLAIR hyperintensities of lentiform nuclei with brightly hyperintense rim surrounding both putamina. (b) T1 weighted MRI: hypointensity over bilateral basal ganglia. (c) Diffusion weighted imaging: high signal intensities in bilateral putamen and globus pallidus and low signal intensities of the forks. (d) Apparent Diffusion Coefficient map: low signal intensities in bilateral putamen and globus pallidus and high signal intensities of the forks suggestive of diffusion restriction and facilitated diffusion respectively**

from the anterior end of the putamen to the stem; (2) the stem, created by merging edematous external and internal capsules at the infero-posterior end of the putamen; (3) the medial arm extending from the stem anteriorly up to one third of the medial edge where it splits into two slightly less T2/FLAIR hyperintense branches engulfing the globus pallidus.<sup>[1]</sup> These two branches are constituted by the edematous medullary laminae, which divide the lentiform nucleus into three masses (the putamen, globus pallidus interna, and externa). Metabolic acidosis was proposed as a key factor in the pathogenesis of this sign in several clinical settings. In cases with uremia, this finding has been reported in patients either not on dialysis or on irregular dialysis and in a child with dialysis disequilibrium during the first dialysis session.<sup>[4-6]</sup> However, in our patient, the clinical and radiological findings appeared late that is after several sessions of hemodialysis. Also, at the point of onset of signs and symptoms there was no significant metabolic acidosis. However, our patient had significant lung infection and multiple episodes of hypotension during hemodialysis sessions. Hence, we postulate that infection, hypoxia and hypotension are possible etiological factors for development of lentiform fork sign in our patient. Hence, lentiform fork sign may be seen in patients even several days after initiation of dialysis and careful attention to acidosis, hemodynamic instability, and treatment of co-existent infection can lead to gradual neurologic recovery and abatement of radiological features.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

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### References

1. Kumar G, Goyal MK. Lentiform fork sign: A unique MRI picture. Is metabolic acidosis responsible? *Clin Neurol Neurosurg* 2010;112:805-12.
2. Grasso D, Borreggine C, Perfett F, Bertozzi V, Trivisano M, Specchio LM, *et al.* Lentiform fork sign: A Magnetic resonance finding in a case of acute metabolic acidosis. *Neuroradiol J* 2014;27:288-92.
3. BeltzEE, Mullins ME. Radiological reasoning: Hyperintensity of the basal ganglia and cortex on FLAIR and diffusion-weighted imaging. *Am J Roentgenol* 2010;195:S1-8.
4. Wang HC, Cheng SJ. The syndrome of acute bilateral basal ganglia lesions in diabetic uremic patients. *J Neurol* 2003;250:948-55.
5. Rathi M, Mudrabetu C. Lentiform fork sign in a case of end-stage renal disease. *Kidney Int* 2012;82:365.
6. da Rocha AJ1, Maia AC Jr, da Silva CJ, Sachetti SB. Lentiform fork sign in a child with dialysis disequilibrium syndrome: A transient MRI pattern which emphasizes neurologic consequence of metabolic acidosis. *Clin Neurol Neurosurg* 2013;115:790-2.

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<b>Quick Response Code:</b> 	<b>Website:</b> <a href="http://www.indianjnephrol.org">www.indianjnephrol.org</a>
	<b>DOI:</b> 10.4103/ijn.IJN_4_20

**How to cite this article:** Radhakrishnan RC, Uthup S, Rajesh L, Pulinilkunnathil RG. Lentiform fork sign in a child on hemodialysis. *Indian J Nephrol* 2021;31:329-30.

**Received:** 25-01-2020; **Revised:** 25-05-2020; **Accepted:** 17-07-2020; **Published:** 07-11-2020.

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