RICH depuis la lettre

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It has been recognised for at least 15 years that there is a category of soft-tissue haemangiomas that are fully grown at birth and involute rapidly without treatment [1]. The diagnostic term “rapidly involuting congenital haemangioma” (RICH¹) was first used 9 years ago [2], at which time it was known that this lesion occurs in the liver [3].

Two papers published in *Pediatric Radiology* cast further light on the important entity of hepatic RICH. First, Franchi-Abella and her colleagues [4] show that many, but not all, antenatally diagnosed liver haemangiomas behave like RICH. In a second paper, my colleagues and I demonstrate the imaging features of biopsy-proven hepatic RICH [5]. We also show in a literature review that many of the cases published as solitary hepatic infantile haemangioma (often called haemangioendothelioma) were almost certainly actually RICH. That some of these patients with RICH *avant la lettre*² were in retrospect treated inappropriately is completely understandable.

What should we make of the observation that patients with RICH are apparently still being treated with hepatic artery ligation [6], resection [7–9] and even transplantation [10]? Why do these reports not recognise RICH as an entity, long after its description in the literature (*depuis la lettre*)?

¹In English, the acronym RICH is usually pronounced *riche* in acknowledgement of the pioneering work of our francophone colleagues in this field. Most English speakers do not adopt a fully French pronunciation, however, because using a guttural rhotic for /r/ makes them sound unacceptably pretentious. Luckily for us, French speakers themselves are somewhat inconsistent in their pronunciation of this phoneme.

²A French expression meaning “before it was described” (literally “before the letter”).

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I think the main reason is that the doctors who look after babies with liver tumours, mostly paediatric hepatologists, liver surgeons, neonatal intensive care specialists and paediatric oncologists, are disconnected from the experts in vascular tumours, who are mostly dermatologists, radiologists and specialists in numerous other fields with an interest in vascular anomalies.

Children with symptomatic hepatic RICH should be treated with aggressive supportive therapy, with embolisation reserved for those who develop uncontrollable cardiac failure [5, 11]. Corticosteroid therapy has not been shown to be beneficial in RICH, and may cause serious complications [4]. Resection of half of a baby’s liver or transplantation is rarely appropriate for this self-limiting condition, may cause fatal complications [7, 10] and should be regarded as a last resort when all other treatments, including embolisation, have failed [12–14]. (This overtreatment of RICH of the liver is nicely described by the French proverb *le médecin est plus à craindre que la maladie*.³)

In these circumstances, the role of paediatric radiologists in diagnosing hepatic RICH and helping other clinicians reach the appropriate management plan may be critical. The radiologist may be the only member of the multidisciplinary team (or tumour board) who is familiar with this condition, and should not be afraid to speak out in the interests of the child.

Remember, for most babies with RICH *l’art de la médecine consiste en amusant le patient tandis que la nature traite la maladie*.⁴

³Literally translated, this means “the doctor is to be feared more than the illness”; in English, we might say “the cure is worse than the disease.”

⁴This aphorism comes from Voltaire (François-Marie Arouet, 1694 – 1778) and may be translated as “the art of medicine consists of distracting the patient while nature treats the disease.”

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