Cardiology in the Young

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Hypertension masquerading as Pediatric Cardiomyopathy: an exercise in cognitive biases

Asim Al Balushi[1,2], Chentel Cunningham[2,3], Manjula Gowrishankar[4], Jennifer Conway[1,2] and Michael Khoury[1,2]

[1]Division of Pediatric Cardiology, Department of Pediatrics, University of Alberta, Edmonton, Canada; [2]Division of Pediatric Cardiology, Department of Pediatrics, Stollery Children's Hospital, Edmonton, Alberta, Canada; [3]Faculty of Nursing, Department of Pediatrics, University of Alberta, Edmonton, Canada and [4]Division of Pediatric Nephrology, Department of Pediatrics, University of Alberta, Edmonton, Canada

Brief Report

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Author for correspondence:
M. Khoury, MD FRCPC, Pediatric Cardiology, Department of Pediatrics, Stollery Children's Hospital, 8440-112th St. NW, Edmonton, Alberta T6G 2B7, Canada. Tel: (780) 407-8361. Fax: (780) 407-3954.
E-mail: Khoury1@ualberta.ca

Abstract

Heuristics and cognitive biases constantly influence clinical decision-making and often facilitate judgements under uncertainty. They can frequently, however, lead to diagnostic errors and adverse outcomes, particularly when considering rare disease processes that have common, masquerading presentations. Herein, we present two such cases of newborn infants with hypertensive renal disorders that were initially thought to have cardiomyopathy.

Heuristics and cognitive biases are regularly present in clinical practice and facilitate judgements under uncertainty. They can, however, result in diagnostic errors and adverse patient outcomes.[1,2] Herein, we present two recent cases of infants presenting with cardiac phenotypes suggestive of hypertrophic and dilated cardiomyopathy.

Case 1

A 6-week-old female was born following an unremarkable pregnancy. During a short neonatal ICU admission for transient tachypnoea of the newborn and hypoglycaemia, an abdominal mass was palpated. A subsequent ultrasound reported a horseshoe kidney. Hyponatraemia was noted and perceived as dilutional with gradual improvement at discharge. Following discharge, the infant had poor feeding and weight gain. Aside from consanguinity, the family history was unremarkable. A murmur was auscultated and a community echocardiogram reported hypertrophic cardiomyopathy. The patient was then referred to our Pediatric Heart Function/Cardiomyopathy clinic.

The patient was irritable throughout her assessment. Her weight was 4.23 kg (birth weight 4.1 kg) at 6 weeks of life. The right arm blood pressure obtained was 99/76 mmHg after multiple attempts. A grade 2/6 systolic ejection murmur was evident. The previously reported abdominal mass was not palpable. An echocardiogram showed concentric left ventricular hypertrophy (z score 3.3–3.5), normal systolic function, and evidence of diastolic dysfunction (Fig 1a, Video). The patient was admitted for further monitoring and evaluation of left ventricular hypertrophy and poor growth.

Laboratory investigations demonstrated hyponatreamia (126 mmol/L). Shortly after admission, the bedside nurse struggled to obtain a blood pressure using an automatic oscillometric device due to continued irritability. A Doppler probe was then used and yielded a systolic blood pressure of 200 mmHg in all four limbs. The patient was urgently transferred to the paediatric cardiac ICU for invasive blood pressure monitoring and treatment. Multiple intravenous antihypertensives were required to obtain blood pressure control. An abdominal ultrasound was repeated revealing autosomal recessive polycystic kidney disease, with confirmation by MRI. A repeat echocardiogram after 2 months of reasonable BP control showed normalisation of the left ventricular hypertrophy (z score 0.4–1.3) (Fig 1b, Video).

Case 2

A 4-week-old male was born at term gestation with an unremarkable pregnancy and birth weight of 3.9 kg. The infant was discharged after 1 day with no concerns. At 3 weeks, tachypnoea was noted. A chest X-ray demonstrated cardiomegaly with increased pulmonary vascular markings, prompting referral to an outside practitioner for an emergent assessment, and an echocardiogram suggesting dilated cardiomyopathy. The infant was urgently referred to our Heart Function/Cardiomyopathy service and subsequently admitted for evaluation.

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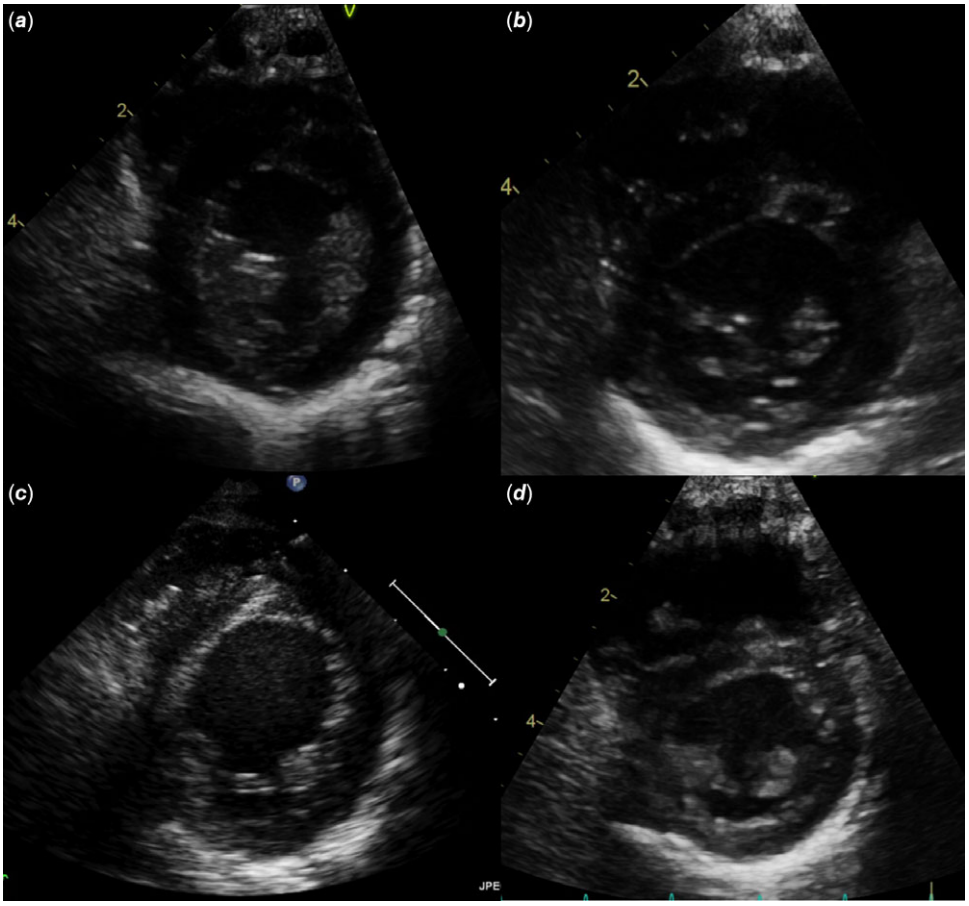

Figure 1. Parasternal short-axis images at end-diastole at the level of the papillary muscles. (***a***) Case 1 baseline assessment demonstrating concentric left ventricular hypertrophy. (***b***) Following adequate hypertension control, normalisation of LVH occurred. (***c***) Case 2 baseline assessment demonstrating mild left ventricular dilation with reduced systolic function. (***d***) Following adequate hypertension control, there was normalisation of left ventricular dimension and systolic function. See Online Video for echocardiographic loops corresponding with these images.

The weight on admission was 4.59 kg. Nursing assessment that evening demonstrated systolic blood pressures of 80–140 mmHg and diastolic blood pressures of 80–100 mmHg. Due to difficulty in obtaining blood pressures, and inconsistency of values obtained (some of which were in a near-normal range), the medical team was not immediately notified of the results. A repeat echocardiogram on admission at our centre demonstrated a mildly dilated left ventricle (end-diastolic dimension z score 2.5) unrestrictive atrial septal defect, moderate mitral valve regurgitation, and moderately reduced systolic dysfunction (ejection fraction 36%) (Fig 1c, Video).

On the day following his admission, the blood pressures remained elevated (systolic BPs 140–150 seconds) and were confirmed using Doppler assessments by the medical team. The patient was subsequently transferred to the paediatric cardiac ICU for invasive blood pressure monitoring and initiation of antihypertensives. An abdominal ultrasound showed a large left renal mass. Surgical nephrectomy and subsequent biopsy demonstrated a stage 2 Wilms' tumor. Adjunctive chemotherapy was employed. Blood pressures quickly normalised in the post-operative period with captopril therapy. Follow-up echocardiogram 1 month later demonstrated normalisation of left ventricular diameter (z score 0.7) and function (ejection fraction 63%) (Fig 1d, Video).

Discussion

The echocardiographic findings in the cases presented represent target organ damage secondary to severe hypertension, a known physiologic adaption to increased arterial blood pressure.[3] Hypertension and hyponatraemia are present in about one-quarter of patients with autosomal recessive polycystic kidney disease at initial presentation.[4] Similarly, hypertension is a common manifestation of Wilms' tumor.[5]

Several cognitive biases contributed to the delayed diagnoses in both presenting cases. Directly referring the patients to the Pediatric Cardiomyopathy clinic with outside diagnoses of suspected cardiomyopathy, resulted in a framing bias, where clinical impressions and decisions were influenced by the circumstances by which the patient presented.[2] The concentric left ventricular hypertrophy, poor growth, and consanguinity in Case 1 contributed to the development of "momentum" in hypothesising that the cardiomyopathy may be due to a metabolic or genetic aetiology. In both cases, the initial hypertensive blood pressure measures were attributed to external factors such as patient irritability and known difficulties in obtaining accurate blood pressures in infancy. This is an example of anchoring bias; an over-reliance on initial information received with insufficient adjustments made to subsequent details.[1,2] The anchoring described resulted in continued misattribution of difficulties in obtaining subsequent blood pressures to technical difficulties, rather than the presence of severe hypertension. Thus, framing and anchoring biases, in addition to the initial diagnostic error on the outside abdominal ultrasound in Case 1, resulted in a delayed diagnosis both of the severe hypertensive urgencies and their underlying renal causes.

Cognitive biases contribute to diagnostic errors and/or suboptimal patient outcomes.[1,2] The present cases provide

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real-world examples of how such near misses and adverse occurrences may unfold.

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Ethical standards. Research ethics board approval was not sought for this case report, though verbal consent was received from the parents of both cases.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951121000093.

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