Reminder of important clinical lesson

How important it is to determine the blood pressure in paediatric patients?

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The prevalence of hypertension among the paediatric population is 1%–2%. The emergency physician should recognise potentially harmful blood pressure (BP) levels and ensure they are adequately treated, in order to avoid life-threatening complications. A hypertensive emergency is a severely elevated BP complicated by target organ dysfunction (cardiovascular, cerebrovascular and/or renal). Hypertensive urgency, however, is a severe elevation in BP without target organ dysfunction. This distinction is critical for the clinical approach. The authors present a case of a severe hypertension due to primary focal segmental glomerulosclerosis. In this case, the lack of BP measurement in the infant surveillance and the devaluation of an albuminuria detected in a previous routine urine examination, have culminated in a late diagnosis of a severe hypertension, with subsequent effects on target organs.

BACKGROUND

The prevalence of hypertension among the paediatric population is 1%–2%. Although the overall prevalence has remained constant for the past decade, there has been a notable increase in the incidence of hypertension in children, likely due to the rising trend toward childhood obesity.¹ The clinical perspective of blood pressure (BP) in paediatric patients has changed in recent years as a consequence of BP regular assessment and a better understanding of normal BP values according to growth and development. This enables the detection of hypertension in children both in the presence of clinical symptoms and in their absence. In addition to detecting hypertensive children requiring therapeutic intervention, the goal of BP measurement in paediatrics is to identify those children or adolescents susceptible of developing hypertension in adulthood. The emergency physician should recognise potentially harmful BP levels and ensure they are adequately treated, in order to avoid life-threatening complications.² A hypertensive emergency is a severely elevated BP complicated by target organ dysfunction (cardiovascular, cerebrovascular, and/or renal). Hypertensive urgency, however, is a severe elevation in BP without target organ dysfunction. This distinction is critical for the clinical approach.

In the case presented, the lack of BP measurement in the infant surveillance and the devaluation of an albuminuria detected in a previous routine urine examination, have culminated in a late diagnosis of a severe hypertension, with subsequent effects on target organs. This case reaffirms the relevance of the routine assessment of BP for early hypertension detection and treatment, and subsequent reduction of both its morbidity and mortality.

CASE PRESENTATION

A 15-year-old Caucasian female presented to the emergency department with worsening headache within the last 48 h.

During the previous 4 months she had pulsatile headache, with occasional vomiting, preceded by nausea, being asymptomatic in the intercrisis period. In her medical history, at 8 years old, she had a routine urine examination with isolated albuminuria, with normal renal function, without subsequent controls. Her family history included a cousin with terminal kidney failure presenting at a young age.

On emergency department first assessment, her BP was 200/147 mm Hg (greater than the 99th percentile corrected for age, gender and height, plus 5 mm Hg). Fundoscopic examination, showed a mild hypertensive retinopathy, without papilledema. The remaining physical examination was irrelevant.

INVESTIGATIONS

Laboratory studies revealed: mild renal insufficiency (glomerular filtration rate of 58.67 ml/min/1.73 m²), nephrotic proteinuria (244 mg/m²/h), hypoalbuminaemia (2.3 g/dl) and dyslipidaemia (total cholesterol of 282 mg/dl, low-density lipoprotein of 181 mg/dl). ECG revealed left ventricular hypertrophy by voltage criteria. Renal doppler-ultrasound excluded vascular stenosis or thrombosis, and showed reduction in the normal parenchyma-sinus differentiation.

OUTCOME AND FOLLOW-UP

The patient was admitted to complete aetiological investigation and for treatment. She started sublingual nifedipine, associated with enalapril. BP control was achieved with a combination of an ACE inhibitor and an angiotensin II receptor blocker (Losartan). The renal biopsy confirmed a chronic nephropathy, revealing signs of primary focal segmental glomerulosclerosis (FSGS). Once the diagnosis was established through renal biopsy, potential secondary causes that would require specific therapeutic approaches were ruled out. Based on the possibility of a genetic

aetiology, immunosuppression was not instituted and she presented a progressive reduction of proteinuria. After 2 years of follow-up, she maintains stable BP and renal function, and a subnephrotic proteinuria. The genetic test for NPHS2 was negative.

DISCUSSION

The systolic and diastolic BP increases progressively over time, parallel to the growth and body development, presenting different patterns according to gender.

In the adult population, hypertension is an important cardiovascular risk factor. However, this association remains controversial in the paediatric population since the development of cardiovascular complications is the result of a combination of age and other risk factors. Subsequently, the definition of normal values in children and adolescents is based in the BP percentile distribution, and hypertension is a statistical diagnosis.^{1–3} According to the criteria of the fourth report,³ hypertension in children and adolescents is defined as sustained systolic and/or diastolic BP elevation greater than or equal to the 95th percentile for age, gender and height, measured on at least three separate occasions with the auscultatory method. Ambulatory BP measurement is now increasingly recognised as being indispensable to the diagnosis and management of hypertension. The incorporation of BP measurement into routine paediatric healthcare should enable the detection of asymptomatic hypertension. Once hypertension is confirmed, underlying aetiology, end-organ damage and cardiovascular risk factors should be assessed.^{2–3} Acute severe hypertension has traditionally been divided into hypertensive emergencies and hypertensive urgencies. In an age of widespread hypertension screening and treatment, the role of chronic, progressive hypertension as a cause of hypertensive emergency is still difficult to clarify. In few cases, a previously unrecognised form of secondary hypertension may be the underlying cause and requires early recognition. Unfortunately, the recognition and treatment of hypertension in the general population are still not adequate, and many physicians may be complacent regarding the need for aggressive control of elevated BP. Thus, many patients with unrecognised or undertreated hypertension may be at increased risk of a hypertensive emergency, as well as subjected to its more insidious consequences. In the case presented, the lack of previous BP assessment in the child surveillance resulted in a delayed diagnosis of hypertension, with progression to a hypertensive urgency. Routine BP screening is recommended for all otherwise healthy children aged 3 years and older, at least once a year, in a medical setting. BP measurements also should be performed in ill and potentially symptomatic children and children younger than 3 years who are believed to be at increased risk for hypertension due to coexisting medical conditions.^{4–5}

Management of patients with acute hypertensive syndromes should be tailored to each individual case and based on the absolute value of BP, and on the presence or absence of target organ dysfunction. Hypertensive urgencies can be treated with oral antihypertensive agents and, after a period

of monitoring, the patient can safely be discharged with close outpatient follow-up and adjustment of therapy. A patient presenting with a hypertensive emergency should be admitted to an intensive-care unit. Intravenous administration of antihypertensive drugs is generally preferred. In general, most extra-cerebral end-organ dysfunction benefits from a rapid decrease of BP. It is recommended that BP should be reduced by no more than 25% within the first 2 h, then reduced gradually to normal values over the next 3 to 4 days. This happens because in long-standing hypertension, cerebral vascular auto-regulatory mechanisms are frequently overwhelmed and can no longer compensate for rapid changes in pressure. Rapid lowering of BP in such cases can result in serious neurologic sequelae.^{2–4} Medications used to treat hypertension should be chosen according to their side-effect profile, availability and physician familiarity with the drug. Much controversy has arisen in the literature over the use of nifedipine in the pediatric population.^{3–7–9} In adults, reports of adverse cardiac and neurologic sequelae secondary to hypotension from the use of nifedipine have led to a moratorium on its use. Similar sequelae related to hypotension have not been observed in children, with case reports being published detailing rebound hypertension causing adverse neurologic events associated with the use of short-acting nifedipine. Nevertheless, many nephrologists continue to use nifedipine for the treatment of moderate to severe hypertension,² as in the case presented. Blaszk *et al*⁶ published a retrospective study of 117 children treated with nifedipine, determining that the drug is safe and effective provided the initial dose is no greater than 0.25 mg/kg. No significant side effects were observed during the study, as in this case. However, due to the unpredictable nature of the orally administered drug, it is recommended that use of nifedipine should be limited to hypertensive urgencies. Intravenous medications are better suited for hypertensive emergencies.

We present a case of an urgency hypertension, secondary to primary FSGS. Proteinuria is a defining feature of FSGS, and, approximately 75%–90% of children have the nephrotic syndrome at presentation. A subset of patients may have asymptomatic proteinuria noted on routine urinalyses, as in the case presented. FSGS is a group of clinical-pathologic syndromes sharing a common glomerular lesion and mediated by diverse insults directed to or inherent within the podocyte.¹⁰ The natural history of FSGS is variable reflecting the pathologic diversity of glomerular lesions. The specific treatment approach is still empirical, and no consensus has evolved because of a lack of prospective controlled trials. The goal of therapy is to induce a complete or partial remission of proteinuria and preserve renal function. Even partial remission is associated with improved long-term survival.^{9–10} The treatment of primary FSGS is empiric and based on the rationale that the permeability factor derives from unregulated immune response. There is no evidence to support glucocorticoid therapy in adaptive or genetic forms of the disease. Some genetic forms may respond to empirical therapy with calcineurin inhibitors.¹⁰

Learning points

- ▶ Routine BP screening is recommended for all otherwise healthy children aged 3 years and older, at least once a year, in a medical setting. BP measurements also should be performed in ill and potentially symptomatic children and children younger than 3 years who are believed to be at increased risk for hypertension due to coexisting medical conditions.
- ▶ The emergency physician should recognise potentially harmful BP levels and ensure they are adequately treated, in order to avoid life-threatening complications. The distinction between hypertensive emergency and hypertensive urgency is critical for the clinical approach.
- ▶ Most childhood hypertension, particularly in preadolescents, is secondary to an underlying disorder. Renal parenchymal disease is the most common cause of hypertension.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Pinto Pais I, Novo A, Figueiredo M, Mota C. How important it is to determine the blood pressure in paediatric patients? *BMJ Case Reports* 2012;10.1136/bcr-2012-006186, Published XXX

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