Aortic coarctation suspected by Doppler echocardiography of renal arteries in hypertensive patients referred to a hospital outpatient hypertension clinic

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Coarctation of the aorta is the fourth most frequent form of congenital cardiovascular disease, which is diagnosed by the presence of higher blood pressures in the arms than in the legs. In this report we describe 3 cases of aortic coarctation, in which the correct diagnosis was suspected only months or years after the detection of hypertension, when a renal ultrasound examination was requested, despite the fact that the hallmarks of the disease were present at the physical examination in all patients.

A marked reduction in renal flow velocities was suggestive of proximal aortic stenosis in all 3 cases.

We conclude that the diagnosis of aortic coarctation, an uncommon but not so rare form of secondary hypertension, by renal ultrasonography rather than by a complete physical examination, reflects a commitment failure of physicians in everyday management of hypertension.

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Introduction

Major hypertension guidelines recommend that all hypertensive patients should undergo a comprehensive medical history, physical examination and a limited number of routine investigations. The aim of the clinical assessment is to investigate secondary causes of high blood pressure (BP), associated modifiable cardiovascular risk factors, evidence of target organ damage and concomitant diseases or accompanying clinical conditions, all of which may guide the physician in decision-making about lifestyle changes and pharmacologic treatment[1-3].

A specific cause of BP elevation can be identified in a small minority of adult subjects with hypertension. Coarctation of the aorta is the fourth most frequent (7.5%) form of congenital cardiovascular disease[4,5], which is commonly diagnosed by the presence of higher BP in the arms than in the legs. The aim of this paper was to present 3 cases of previously unsuspected aortic coarctation in which the correct diagnosis was made by Doppler ultrasound examination of renal arteries, aiming at excluding renovascular hypertension.

Description of cases

Case 1. A 34-year-old woman was sent to our attention because antihypertensive treatment with enalapril 20 mg/day plus hydrochlorothiazide 12.5 mg/day, later substituted by enalapril 20 mg plus doxazosin 4 mg/day, had failed to normalize her BP values. Two years before, her family physician had prescribed full laboratory examinations: renal function and serum electrolytes were found normal, fundoscopy had shown diffuse arteriolar narrowing and an electrocardiogram had revealed a typical Wolff-Parkinson-White pattern. The patient was a social drinker, smoked about 5 cigarettes a day since the age of 17, had a sedentary lifestyle and denied assumption of licorice, steroids or oral contraceptives.

Doppler scanning of the renal arteries showed abnormally low systolic and diastolic peak velocities, both markers of diffuse hypoperfusion, at the proximal level of main renal arteries (near the abdominal aorta), at the ilar level and distally in intraparenchymal arterioles, bilaterally (Fig. 1). Echocardiography showed a moderate concentric left ventricular hypertrophy: left ventricular mass index 146 g/m^2 (normal

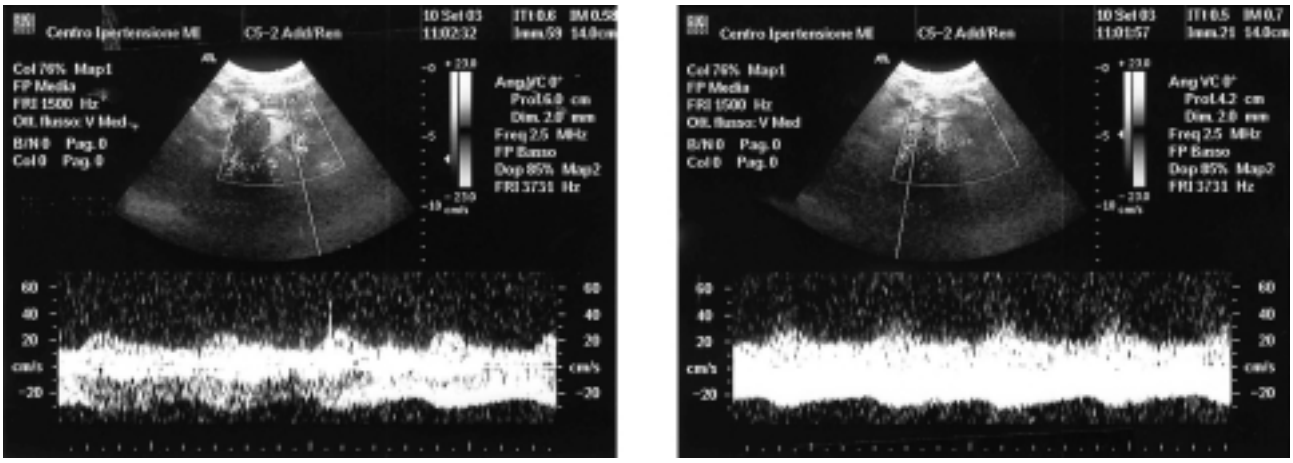

Figure 1. *Doppler examination of the renal arteries, recorded at the proximal level (near the aorta), shows slowed systolic upstroke and decreased systolic peak velocity bilaterally.* Left panel: *left renal artery;* right panel: *right renal artery.*

value < 110 g/m^2) with a relative wall thickness of 0.46 (normal value < 0.45) and a bicuspid aortic valve, but failed to obtain a good visualization of the site of the coarctation or an abnormal high velocity systolic jet in the aortic arch.

At physical examination a 2/6 midsystolic murmur was heard over the mesocardium as well as over the interscapular area; arterial pulses at both lower extremities were reduced.

Computed tomography showed a marked isthmic coarctation (1.2 cm) with relevant post-stenotic vessel dilation (Fig. 2). The coarctation was treated successfully with reconstructive surgery (end-to-end anastomosis). Echo-Doppler ultrasound of the renal arteries, performed after surgery, showed normal renal blood flow patterns bilaterally (Fig. 3).

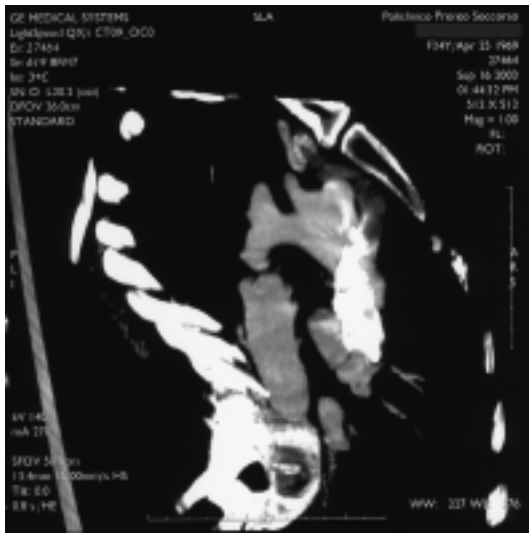

Figure 2. *Computed tomography demonstrates a severe stenosis of the descending aorta distal to the left subclavian artery.*

Case 2. An asymptomatic 14-year-old boy with positive family history for hypertension was referred to our

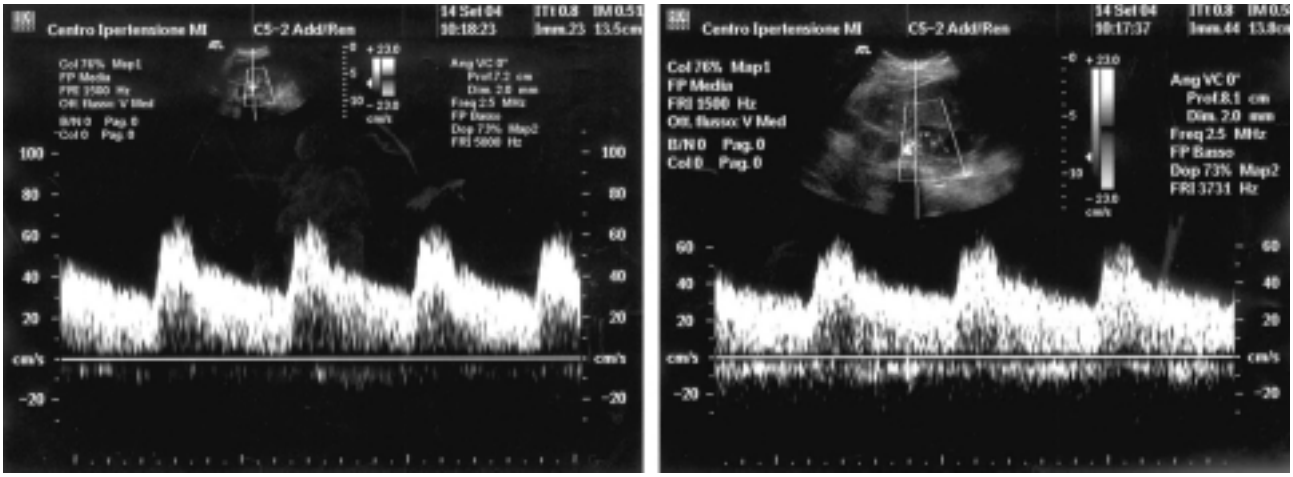

Figure 3. *Doppler examination of the renal arteries, performed after reconstructive surgery of aortic coarctation, shows normal blood flow patterns bilaterally.* Left panel: *left renal artery;* right panel: *right renal artery.*

hospital outpatient clinic for a suspicion of renovascular hypertension with the request of a renal ultrasonographic examination. The boy had been regularly practicing sports (soccer and basketball) in the last few years. Three months earlier BP values of 145-150/95-100 mmHg were found at casual measurements. At that time, his physician had performed diagnostic procedures including routine blood chemistry and urinalysis, electrocardiogram, echocardiogram, plasma renin activity, plasma aldosterone and 24-hour ambulatory BP monitoring. All these examinations gave normal results, except ambulatory BP monitoring that confirmed high BP values (average day-time BP 147/98 mmHg), with an abnormal circadian profile characterized by reduced BP fall during night-time.

Renal ultrasonography showed two kidneys of normal size, but Doppler examination revealed a slowed systolic upstroke and reduced peak systolic velocities in both main renal arteries and in intrarenal arteries. A similar spectral trace was recorded from the abdominal aorta.

The ultrasonographic diagnosis of aortic coarctation was confirmed by previously undetected low intensity ejection heart murmur and a marked reduction in arterial pulses in both lower extremities. Spiral computed tomography showed a severe isthmus coarctation.

Case 3. A 32-year-old woman was referred by her family doctor to our hospital outpatient clinic for investigating the possibility of renovascular hypertension since a few weeks earlier elevated supine plasma renin activity (4.2 pg/ml/hour, normal value < 2.0 pg/ml/hour) and plasma aldosterone concentrations (321 pg/ml, normal value < 150 pg/ml) had been found. The patient had no family history of hypertension, ischemic heart disease nor diabetes. Since the diagnosis of hypertension had been made at the age of 27, intake of oral contraceptives had been withdrawn and the patient was regularly treated with amlodipine, without achieving, however, a satisfactory BP control. She was substantially asymptomatic. Renal ultrasonography showed normal size and structure of both kidneys but the Doppler analysis of renal blood flow guided by echo-color imaging revealed a reduced peak velocity in both main renal arteries and abdominal aorta.

In addition to mild left ventricular hypertrophy (left ventricular mass index 130 g/m^2) with an increased relative wall thickness of 0.48, cardiac ultrasound detected a bicuspid aortic valve without aortic insufficiency. Suprasternal examination of the aortic arch revealed a spectral flow pattern with rapid early systolic acceleration and peak systolic velocity of 4.1 m/s, without a significant diastolic run-off.

In this case as well the physical examination revealed reduced femoral, tibial and pedal pulses in both legs.

A subsequent computed tomography confirmed the presence of a severe aortic coarctation which was successfully corrected by surgery, after the first attempt by balloon angioplasty had failed.

Discussion

Coarctation of the aorta is a congenital abnormality, most commonly located distal to the origin of the left subclavian artery near the insertion of the ligamentum arteriosum, and comprises 6-12% of all congenital cardiovascular diseases. More than half the patients have associated cardiac malformations, such as ventricular septal defects, aortic valve stenosis or left-sided hypoplasia. In addition, systematic echocardiographic studies have shown that a bicuspid aortic valve is a common finding being present in about two thirds of the cases (50-85%). Unlike the critically ill patients diagnosed in the neonatal period, the majority of adolescent or adult subjects are asymptomatic or only mildly symptomatic for headaches, leg cramps or muscle weakness. Hypertension is often found in the course of occasional BP measurements and is the main reason why these patients usually come to clinical attention.

In this report we describe 3 cases of aortic coarctation, in which the correct diagnosis was suspected only months or years after the detection of hypertension, when a renal ultrasound examination was requested, despite the fact that the hallmarks of the disease were present at the physical examination in all patients. Our findings can be discussed as follows.

First, hypertension is a major risk factor in the development of cardiovascular disease and poses significant public health problems. Despite the availability of very effective treatments, the control of high BP in the community is far from optimal. The failure of physicians to adequately treat hypertension to goal level and to detect secondary causes of hypertension is a major factor contributing to poor control. The delayed diagnosis of aortic coarctation, and its clinical negative consequences (expensive and inappropriate diagnostic procedures as well as ineffective treatment associated with high risk of cardiovascular complications) was clearly related to an inaccurate initial clinical evaluation. A comprehensive physical examination at the time of presentation of high BP in all 3 patients would have been sufficient to raise the suspect of an aortic coarctation and to plan timely investigations.

Second, our data reflect once again the important limitations regarding the management of hypertension in the "real world". Why is there such a discrepancy between official recommendations of hypertension guidelines and everyday clinical practice? Despite the fact that hypertension guidelines clearly state that in addition to BP measurement, physical examination should search for evidence of additional risk factors, for signs suggesting secondary hypertension, and for evidence of organ damage[2], these indications, as shown in the present report, are often neglected by practitioners. Two possible explanations for this behavior are: 1) insufficient knowledge by too many doctors of hypertension guidelines; 2) limited implementation and compliance of physicians to these guidelines. In particular the sec-

ond issue was analyzed by many authors and ourselves. In a multicenter region-wide study conducted in Italy we explored the general practitioners' compliance to the 1999 World Health Organization/International Society of Hypertension guidelines in the clinical management of newly diagnosed hypertensive patients[6]. The physician's compliance was evaluated by a questionnaire containing questions about diagnostic work-up and treatment made at the time the first diagnosis of hypertension was made. A minimum clinical and laboratory work-up, as suggested by the guidelines, had been carried out only in a minority of patients (10%). Furthermore, physical examination was not fully performed in a large fraction of patients (40%). These findings support the view that most of the errors in the diagnosis, as those reported here, are due to the fact that the clinical assessment is often limited to BP measurement in one arm and followed immediately by drug prescription.

Third, in the case 2 an echocardiographic examination performed before renal ultrasonography failed to show any cardiovascular abnormality; it should be remarked that in this patient aortic coarctation should have been detected or at least suspected if the ultrasound examination had been correctly performed, analyzing cardiac and vascular structures also by the suprasternal approach.

Finally, some aspects concerning the clinical value of renal ultrasonography in detecting aortic coarctation deserve to be mentioned. On Doppler ultrasound, the normal flow pattern in the main renal arteries shows a sharp acceleration in the antegrade velocity with a peak during systole, followed by a rapid decrease. A renal artery stenosis is typically characterized by an increased peak systolic and diastolic velocity at the site of the narrowing, with dampening of the waveforms distally to the obstruction (when it is > 50-60%) associated with a decreased slope of systolic acceleration and diminished systolic peak velocity. A marked reduction in both abdominal and renal flow velocities is suggestive of proximal aortic stenosis; all sonographists should be aware of the clinical relevance of these signs. Previous studies performed in a pediatric setting reported that abnormalities in renal flow such as "tardus-parvus" Doppler signals and/or alterations in abdominal aortic pulsatility are suggestive of aortic coarctation[7,8]. Most of these children, however, unlike our patients, were referred for renal ultrasonography because the clinical hallmarks of the disease were not reliable, as femoral pulses were palpable and lower extremity BP was normal, in relation to the development of luxurious collateral circulation.

We conclude that the diagnosis of aortic coarctation by renal ultrasonography rather than by a complete physical examination reflects a commitment failure of physicians in everyday management of hypertension.

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