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Unusual Vascular Malformation of the Kidney in Both Twins of a Monozygotic Pair: Short Case Report

G. Mastroberardino[1], A. Sciarra[1], R. Gualtieri[1], C. Di Fusco[2], P. Grammatico[2], G. Del Porto[2]

[1] *Fourth Chair of Clinical Medicine, and* [2] *Chair of Medical Genetics, School of Medicine, First University of Rome, Italy*

Abstract. A case of double bilateral renal vessels in both twins of a MZ pair is reported for the first time. Even in non-twins, the anomalies reported so far involved the inferior polar arteries in agreement with the embryological development. The two male twins, examined at the age of 14 years, had simultaneously developed a marked hypertension at the age of 7 years. Zygosity was determined by blood group and HLA analysis and various clinical tests were carried out to diagnose the condition. It is suggested that the anomaly is the result of a genetically induced early block in embryonic development.

Key words: Vascular malformation, Twins, Kidney

A number of cases of family malformation of renal vessels have been reported in the literature [2,4,5]. All of these concern doubling of the lower polar branches but no case of doubling of the upper polar branches has so far been reported to our knowledge.The first case of bilateral doubling of the upper polar branches in both twins of a MZ pair is described in the following.

CASE REPORT

Male twins, Enrico and Massimo, aged 14 years. Diagnosed as MZ based on concordance for HLA system (A30, A32, B14, B49, Bw46, DR1, DRw6), blood groups (B Rh +) and general morphology.

Enrico: Premature birth (36 weeks) by cesarean section; one episode of tachycardia at birth treated with digitalis. Enlarged thymus. At 7 years of age, hypertension was observed; the treatment applied did not yield the desired result. Hospitalized for examination and treatment. Arterial blood pressure higher (180/120 mm Hg) than cotwin and 2nd cardiac tone reinforced on the aortic focus. Renin: clino-and orthostatism 0 ng/ml. Aldosterone: clinostatism 520 ng/ml; orthostatism 640 ng/ml. Fundus oculi: significant signs of arterial sclerosis with accentuation of the axial reflex and unusual signs of a-v cross. ECG: electrical axis diverted to the left; picture of ventricular preexcitation (W.P.W.); ventricular impairment. Echocardiogram: concentric hypertrophy of left ventricle with altered radius-thickness ratio. Kidney arteriography: bilateral presence of doubling of kidney vascular district, consisting of upper polar branches (Figure).

Massimo: Premature birth (36 weeks) by cesarean section. At 20 days, infection of lower urinary tract treated in hospital. At 7 years, hypertension was observed and treated as in cotwin. Hospitalized with cotwin. High blood pressure (160/100 mm Hg) was found. Renin: clino-and orthostatism 0 ng/ml. Aldosterone: clinostatism 580 ng/ml; orthostatism 350 ng/ml. Echocardiogram and kidney arteriography: as in cotwin.

DISCUSSION

The described anomaly of renal vascularization in both twins has an exceptional character. Normally, during the evolution of kidney vascularization, renal arteries are obliterated in the caudo-cranial direction until only the definitive renal artery is left [6]. The most common anomaly is the persistence of one or more caudal arteries [1,3,7]. The presence in our twins of supranumerary arteries represented by branches of the upper renal pole is the first reported of its kind. This anomaly was responsible for the clinical features of our twins and became apparent, at different times, within the first 14 years of life. Already the history shows hypertension to have become more accentuated in Enrico at an earlier age; attention to the hypertension of Massimo was drawn by the hypertensive state of his cotwin. The clinical status we found confirms the different evolution of hypertension in the twins. In fact, the sclerosis of the retinal arteries in Enrico and the concentric hypertrophy of the left ventricular wall suggest that the hypertensive state in this twin was not only of longer standing but also more severe than in the cotwin. These clinical findings were further confirmed by the renin and aldosterone blood levels. On the other hand, objective data and laboratory findings in Massimo, while confirming the substantial identity of the malformation, indicated a less advanced clinical condition, but in our opinion an analogous evolution is to be expected unless appropriate treatment is applied.

Special consideration should be given to the W.P.W. anomaly clinically manifested in Enrico already at birth, with an episode of supraventricular paroxysmal tachycardia, the electrocardiographic expression of which could be sporadically observed during hospitalization. The substantial similarity of the malformative pathology, even if apparently dissimilar in its clinical evolution, would seem to point to an etiology of a multifactorial type. In fact, the identical genetic background of the two twins exposed to a different action of environmental factors implies a different clinical evolution with time. In conclu-

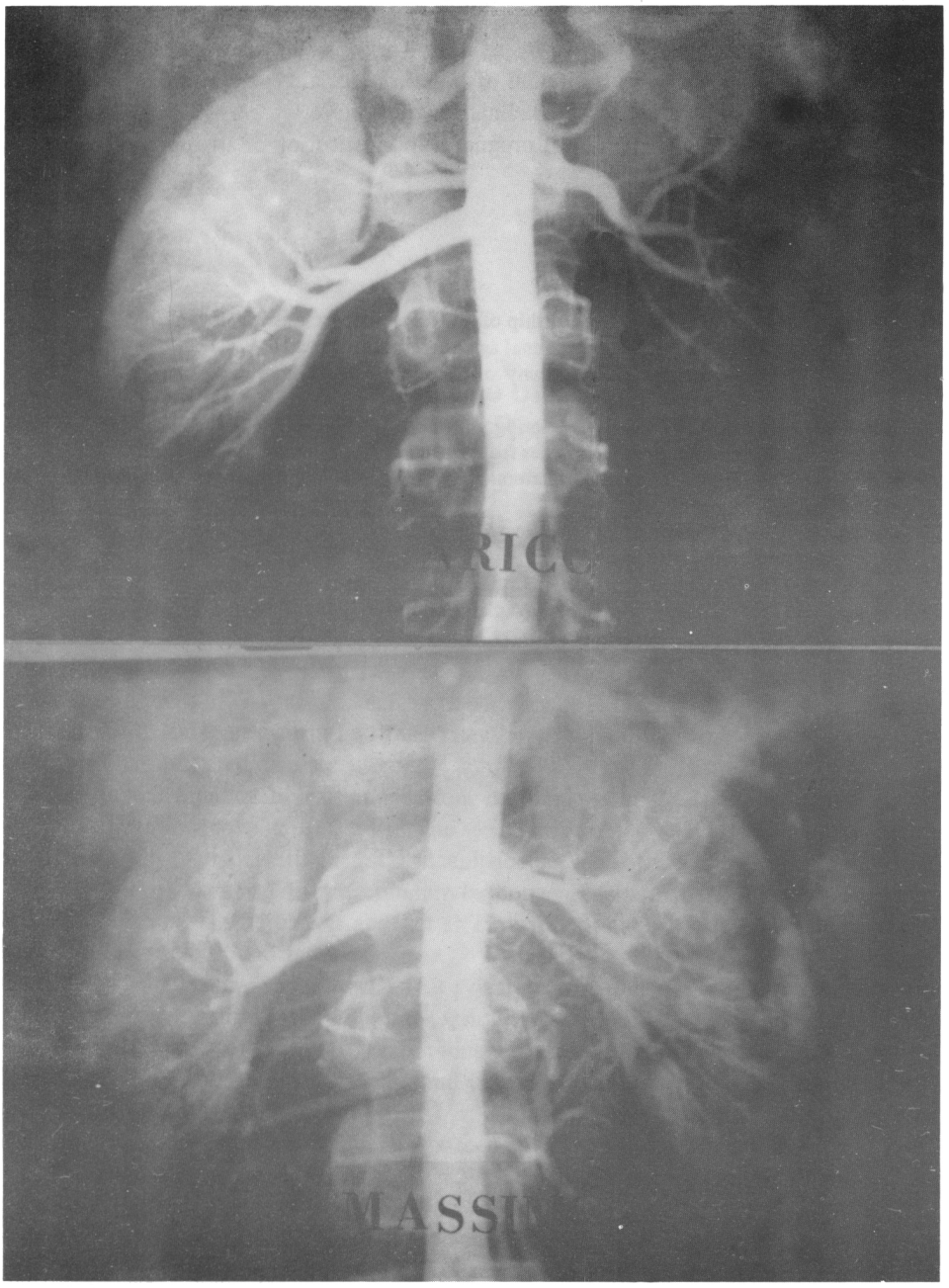

Figure - Arteriography of the two twins: double bilateral renal district in both.

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sion the interest of this case can be summarized as follows:

1) Report of a malformation concerning an upper polar branch never described in the literature until now;
2) Identification of a hereditary model of a multifactorial type, a similar malformation in MZ twins having had a different clinical evolution;
3) Interest from the point of view of prognosis and institution of an appropriate therapy in the clinical evolution.

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Correspondence: Prof. Giuseppe Del Porto, Via San Calepodio 7, 00152 Rome, Italy.