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Situs Viscerum Specularis in Monozygotic Twins

L. Gedda¹, A. Sciacca², G. Brenci¹, S. Villatico², G. Bonanni¹, N. Gueli²,
C. Talone¹

¹The Gregor Mendel Institute of Medical Genetics and Twin Research, Rome; and

²Institute of Medical Pathology, First University of Rome

Abstract. A case is reported of a 14-year old male MZ twin pair, with only one partner affected by situs viscerum inversus, whereby the condition is termed situs viscerum specularis. The high degree of mirror imaging is seen as an abnormal variation of the biological time of the cleavage giving rise to MZ twinning. Electrocardiographic findings are presented and the biological implications of the condition are discussed.

Key words: Situs viscerum inversus, Monozygotic twinning, Laterality, Atriogram

When in a single-born uneven viscera are shifted from the normal to the opposite side of the body, the anomaly is called "situs viscerum inversus". We have observed a pair of MZ male twins only one of whom is affected by this anomaly, so that we have deemed it useful to place a diagnosis of "situs viscerum specularis" and to present this unusual case report.

Maurizio and Fabrizio (Fig. 1) are now 14 years old and present slight differences of height and weight. Pedigree study indicates that the maternal grandfather was also a twin. In Maurizio, the first-born, the heart is on the right, the liver on the left, and the stomach on the right, while in Fabrizio these viscera are normally located on the opposite side as can be seen from the radiograph (Fig. 2).

No mirror imaging is found in the dermatoglyphics and other nonvisceral traits are normal and concordant. The dextrocardiac twin also suffers from slight hearing loss, following otitis, and enuresis – which the cotwin got over at the age of 4 years. Caries of the permanent dentition are present in both twins, but more so in the dextrocardiac one. The twins are both psychologically retarded and introverted. They only speak to their mother.

The results of electrocardiographic examination, are shown in Fig. 3. It can be noted that when the electrodes are placed normally (left), the ECG of the dextrocardiac twin



Fig. 1

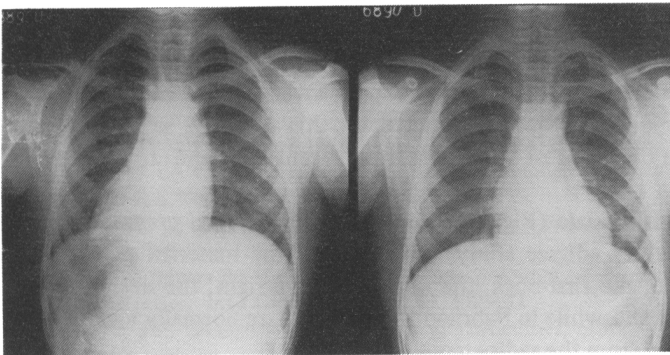
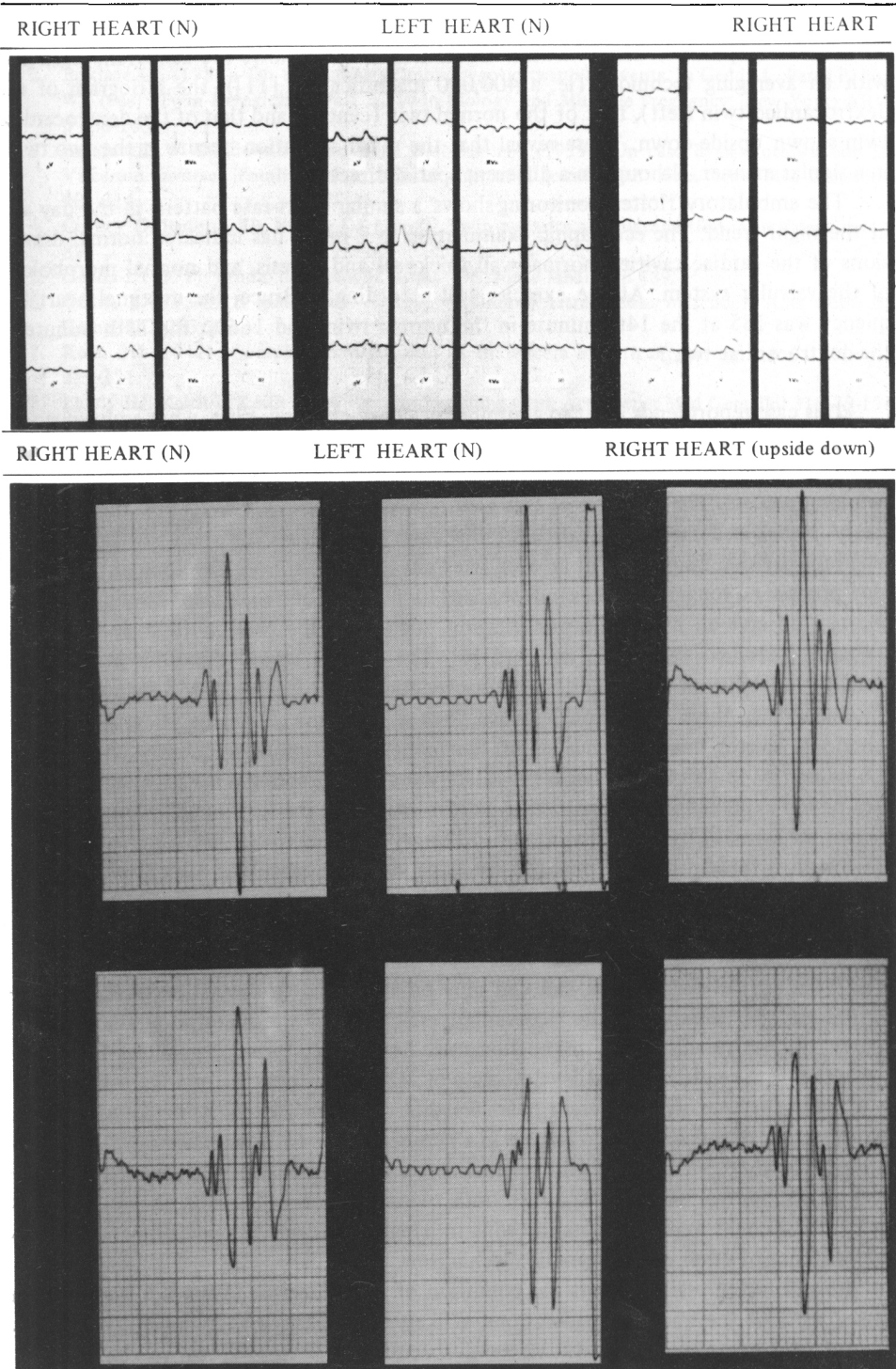


Fig. 2



Figs. 3 and 4 - Electrocardiographic examination of the two twins.

reveals an inversion of polarity in the first derivation and in the six precordial derivations, whereas with an inverted positioning of the electrodes (right), the ECG of the dextrocardiac twin corresponds to that of the cotwin (center). Fig. 4 shows the atriogram registered with an averaging technique (ie, a 400,000 magnification [11]): the atriogram of the dextrocardiac twin (left), that of the normal twin (center), and that of the dextrocardiac twin shown upside-down. These reveal that the atrial activation occurs in the two twins in a similar manner, although in a different spatial direction.

The ambulatory Holter monitoring shows a similar heart-rate pattern in the day and in the night trend. The ecographic examination in B-mode has indicated normal dimensions of the cardiac cavities, normal wall thickness and kinesis, and normal morphology of the valvular system. At the exercise test according to Bruce, the maximal heart frequency was 185 at the 14th minute in the normal twin, and 166 at the 13th minute in the dextrocardiac twin.

This case report lends itself to a number of considerations. First, what embryogenetic mechanism can produce the condition of situs viscerum specularis in MZ twins? We think that this might be related to the timing of action of the genotype underlying the MZ twinning process. The chronon of this genotype is in fact expressed when the first symmetry planes and the lateralization of the embryo contents have already taken place. When the division takes place at an even later phase in the embryo development, the twinning process no longer produces independent individuals, but coalescent cotwins in which one can observe an evident mirror imaging, such as in the case of the male dicephalus tribrachius studied by Gedda in 1954 [6]. The process would therefore involve a secondary variability of the hereditary chronogenetic variant leading to MZ twinning [2].

Second, is there a relation between such an anatomical mirror imaging and the functional mirror imaging, such as left-handedness, that more frequently characterizes MZ twins? [5:pp 489 ff]. We believe it is likely that this functional mirror imaging may in turn be the expression of an anatomical mirror imaging at the level of the nervous system, this too determined by a late division of the embryo leading to MZ twinning.

Third, is there a relation between the situs viscerum specularis of MZ twins and the situs viscerum inversus of a single-born? It may be recalled that, in his classical works on twin anatomy published in Japan back in the 1930s, Tsuchiya [12] had already shown that differences in the heart weight exist in cotwins already in the fetal life and are higher in MZ than in DZ twins. Now, this can only be attributed to the action of endouterine factors and particularly to the differential efficiency of the materno-fetal circulation known to particularly affect monochorionic, hence MZ, twin pregnancies [4]. These diversities in the embryological development of MZ twins, resulting, among other things, in MZ twin fetuses with different heart weights, are in fact likely to be responsible for the early loss of one of the two fetuses in a number of MZ twin pregnancies, and possibly in part for the "Vanishing Twin" phenomenon [8]. Otherwise, the existence of five twin-pairs (four MZ and one DZ) with both twins concordant for situs viscerum inversus [1,3,7,9,10] could imply the loss of twins in a higher multiple pregnancy, or conceivably the existence of other embryogenetic mechanisms.

Finally, when one considers the condition of situs viscerum inversus in a single-born in the light of the above considerations, a likely hypothesis is that this is the surviving twin of a MZ pair affected by situs viscerum specularis.

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Correspondence: Professor Luigi Gedda, The Mendel Institute, Piazza Galeno 5, 00161 Rome, Italy.