Connexions, relations, discordance, and distortions

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It is impossible to discuss complex cardiac anomalies without clearly differentiating between relations and connexions. Relations describe broad spatial interrelationships of two cardiac structures. In contrast, connexions specifically state the fashion in which two structures are connected one to the other irrespective of their relations. Connexions and relations may be, and indeed usually are, the same. Unfortunately this is not always the case, which is precisely why we prefer the concept of connexions. To take two examples; firstly, it is possible for the normally situated right atrium to be normally connected to the morphological right ventricle, but for this right ventricle to be left sided (Anderson, Shinebourne, and Gerlis, 1974). Thus the connexions are normal, but the relations of the ventricles are clearly abnormal, since they are spatially inverted. Secondly, it is usual for the aorta to be posterior and right sided with relation to the pulmonary artery in a patient with situs solitus and normally connected ventricles. However, it has been shown that the normally related aorta in this situation can take origin from the *right* ventricle and not the left (Van Praagh *et al.*, 1971; Wilkinson *et al.*, 1975). Using a relations concept, it is not possible to describe this connexion as transposition. However, unequivocally the ventriculo-arterial connexion is one of transposition (Van Praagh *et al.*, 1971; Wilkinson *et al.*, 1975). It is the intracardiac connexions which are determined by embryogenesis. These in turn determine the haemodynamic condition of the patient and the corrective surgery which may be required. In contrast, relations may be affected by multiple extracardiac events, and by rotation of the heart following completion of its normal development. Thus, we believe that a

nomenclature based upon connexions is both more meaningful and more helpful than one based upon relations. This holds good not only for the embryologist, but for the cardiologist, surgeon, radiologist, and pathologist.

In this issue of the *British Heart Journal*, de la Cruz and her colleagues (1976) propose a nomenclature together with rules for diagnosis which are based entirely upon relationships. If these rules are applied to the two cases quoted above, then diagnoses of, firstly, a discordant atrioventricular relation and, secondly, of normally related great arteries without transposition would be made. Such diagnoses are not only misleading, but could be positively dangerous in a clinical context.

It is also suggested that viscero-atrial situs should be established from hepatic and inferior vena caval position, and ventricular position from examination of the relationships of the infundibula and great arteries. In order to establish the latter relationships, de la Cruz *et al.* (1976) provide complex and lengthy 'rules'. This seems to us to be using a sledgehammer to crack a nut.

Angiocardiographically, ventricles need only be defined in terms of their intrinsic qualities. With the possible exception of coronary arteries, it is actually misleading to refer to other structures. One of us (F.J.M.) has worked in a centre in which, for the past five years, all symptomatic complex 'transpositions' and 'malpositions' of appropriate age have been referred for corrective surgery. Ventricular localization has been carried out solely by examination of the diastolic shape and trabecular pattern after injection of contrast medium into both ventricles, and atrioventricular connexion established by inspection of the atria, catheter course, and atrioventricular valves. No error in these respects has been made in over 1300 cases studied. This experience is not regarded as anything special, but

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as normal for any catheterization laboratory involved in the preoperative assessment of such patients. 'Rules' for predicting ventricular positions are by no means a new proposition. Twelve years ago, an attempt was made at prediction of ventricular position from great arterial position using the 'loop' rule (Van Praagh *et al.*, 1964). This 'rule' was subsequently shown to be inaccurate in about 30 per cent of cases (Carr *et al.*, 1968). To his credit, Van Praagh subsequently admitted its fallibility (Van Praagh, 1972). By contrast, when the 'rules' put forward by de la Cruz and Nadal-Ginard (1972) were shown to have exceptions, a new term (arterio-ventricular discordance) was coined to describe those hearts which broke the rules (de la Cruz *et al.*, 1974). If the 'rules' themselves are confusing, this term is endowed with even more potential to confuse. The term 'discordance' is used with an entirely different meaning from its application to atrioventricular relations (or connexions). In the latter situation, discordance indicates a situation opposite to normal. By analogy, a discordant arterio-ventricular relation (or connexion) should also be the opposite of normal, namely transposition. This is indeed the definition of ventriculo-arterial discordance given by Kirklin and his colleagues (1973). But, according to de la Cruz *et al.* (1974, 1976) arterio-ventricular concordances and discordances can each coexist with either transpositions or crossed (normally related) great arteries.

There are additional fundamental scientific objections to the classifications proposed by de la Cruz and her colleagues (de la Cruz and da Rocha, 1956; de la Cruz and Nadal-Ginard, 1972; de la Cruz *et al.*, 1974, 1976). De la Cruz and Nadal-Ginard (1972) laid down other 'rules' for establishing atrial situs. They defined, as others had done (Van Praagh *et al.*, 1964), situs solitus, situs inversus, and heterotaxias. They then stated that, 'Heterotaxias... due to their anatomic and embryologic features... do not conform to the rules which apply to situs solitus and inversus' (de la Cruz and Nadal-Ginard, 1972). However, they do not indicate how heterotaxias are to be clearly differentiated in life from situs solitus and inversus. We would point out a mid-line liver is a very poor indicant of situs ambiguus (heterotaxia). From the detailed reports of 84 patients with abnormal cardiac situs or position from the Hektoen Research Institute, it is clear that a mid-line liver is the exception rather than the rule in asplenia and polysplenia (Lev *et al.*, 1968, 1971; Liberthson *et al.*, 1973). Had atrial situs been predicted from liver position, it would have been correct in only 37 per cent of cases. In

contrast, lung lobulation would have given the correct prediction in 72 per cent of cases. Bronchial visualization, an easily performed clinical investigative technique, could well have given even better prediction. A more recent study has shown that measurement of bronchial lengths enables clear distinctions to be made between thoracic lateralization (solitus or inversus) and thoracic isomerism (heterotaxy) (Partridge *et al.*, 1975).

Similar divergences in interpretation and opinion are found with regard to 'arterio-ventricular' relations, or, as we would prefer to describe them, ventriculo-arterial connexions. De la Cruz and her colleagues (1956, 1972, 1974, 1976) describe normal relations, transposition and an intermediate group named 'partial distortions' (Angelini and Leachman, 1973). These terms describe *only* relations, so a double outlet ventricle connexion can, according to de la Cruz *et al.* (1976), coexist with any of these relations. We submit that this concept is equally confusing. It has been convincingly shown in recent years, both from angiographic studies (Guerin *et al.*, 1970) and from study of necropsied material (Lev *et al.*, 1971; Goor and Edwards, 1973; Anderson *et al.*, 1974a) that a spectrum of abnormalities exists between transposition on the one hand and double outlet ventricle on the other. When a spectrum exists, it is not possible to make clear-cut distinctions. Instead, an arbitrary distinction is necessary such as that proposed by Kirklin *et al.* (1973) demanding more than one and a half great arteries arising from the same ventricle for the definition of a double outlet ventricle. Van Praagh (1973) has marshalled excellently the arguments against the description 'partial distortion'. We would add that unless specific 'rules' are laid down for the distinction between partial distortions and other trunco-conal morphologies, the 'rules' for arterio-ventricular discordance are valueless, since partial distortions are specifically excluded from these 'rules' (de la Cruz *et al.*, 1976). This point is particularly relevant in the case of anatomically corrected malposition. In order to explain why this does not obey the rules, de la Cruz *et al.* (1974, 1976) state that this is a partial distortion. If the case descriptions to which they refer (Van Praagh and Van Praagh, 1967; Anderson Arnold, and Jones, 1972) are consulted, it is found that in three of four cases the aorta was specifically stated to arise anteriorly to the pulmonary artery. According to de la Cruz's own nomenclature (1974, 1976), this fact would make them 'transpositions' rather than 'partial distortions'. Partial distortion should surely serve a more useful function than constituting a

convenient hiding place for cases which 'break the rules'.

De la Cruz and her colleagues (1976) argue that their hypothesis is based upon embryological facts. It is very difficult to determine what constitutes an embryological 'fact'. We would contend that they refer to 'embryological interpretations'. They discuss their concepts and support them by reference to work of other noted embryologists. When considering contrary views, however, they argue only against the 'conal growth' hypothesis of Van Praagh. If the original argumentation of Van Praagh is read relative to this hypothesis (Paul, Van Praagh, and Van Praagh, 1968), it becomes very evident that the hypothesis was propounded precisely because the classical 'straight septum' hypothesis, which de la Cruz *et al.* (1976) still support, could not account for the anatomy of specimens with transposed arteries. Somewhat ingenuously, de la Cruz *et al.* (1976) fail to mention this point. They also fail to point out that at that time Paul *et al.* (1968) considered a conal *absorption* hypothesis. They did not advance such an hypothesis because they considered it was not supported at that time by embryological investigations, though pointing out that Butler (1952) had shown conal absorption in the chick heart. Paul *et al.* (1968) proposed conal growth because it was supported by *anatomical* fact. De la Cruz *et al.* (1976) suggest that the mechanism of transference of the aorta to the left ventricle is unknown. This is also somewhat ingenuous. The mechanics of this process were elucidated by Pernkopf and Wirtinger (1933) and endorsed by Asami (1969). They were again endorsed by Goor, Dische, and Lillehei (1972). It is surprising that de la Cruz *et al.* (1976) quote the article of Goor as ammunition against Van Praagh *et al.* (1964), but do not comment on Goor's suggestion that transposition results from differential conal absorption coupled with lack of conal inversion. This concept has subsequently received support from the work of Anderson *et al.* (1974b). Above all, de la Cruz *et al.* (1976) do not quote the important paper of Los (1968). His reconstructions in the human convincingly showed that, though the conotruncal ridges formed in spiral fashion, they *fused* in straight fashion. This finding has been confirmed in the chick heart by the investigation of Dor and Corone (1973). Thus in the *normal* heart the conus septum is a straight and not a spiral structure. This, combined with the arguments of Paul *et al.* (1968), plus the embryological studies of Pexieder (1975), convince us that a differential conal *absorption* hypothesis gives a much better explanation of transposition than the 'straight septum' hypothesis

promoted so strongly by de la Cruz *et al.* (1976). However, we do not claim these points as facts; they are interpretations. As such they could be wrong. We would request that de la Cruz and her colleagues (1976) review ALL the evidence against their own hypothesis, rather than attacking a conal growth concept which is no longer propounded by its originator (Van Praagh, 1973).

Beyond doubt, de la Cruz and her colleagues have, over the years, made a major contribution to our knowledge of congenital heart disease, particularly in their meticulous descriptions of malformed hearts. In drawing attention to the more controversial aspects of their work, we wish only to stimulate readers to make their own critical judgement on these issues. There is little point in enumerating the inconsistencies of a particular viewpoint without providing a convincing and more consistent alternative. It is our hope that a nomenclature based upon connexions rather than relations, as proposed elsewhere in this issue (Shinebourne, Macartney, and Anderson, 1976), will do precisely this.

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