Double outlet right ventricle

Study of 27 cases

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Out of 1610 children's hearts with congenital malformations there were 27 specimens showing double outlet right ventricle. Cases with dextrocardia, situs inversus, or l-ventricular loop were excluded. Anatomical examination was performed with particular reference to the infundibular region, the great vessels, and the ventricular septum. The commonest associated malformations were ventricular septal defect and pulmonary stenosis. Aortic stenosis was the predominant finding in those cases dying in the neonatal period. An aortic conus was associated with pulmonary stenosis, ventricular septal defect, and d-transposition, a pulmonary conus with ventricular septal defect and a double conus with stenosis of either great vessel. The anterior vessel always had a muscular conus and the posterior vessel was commonly stenotic.

The term 'double outlet right ventricle' (DORV) signifies that both great arteries arise completely from the morphological right ventricle and that the only outlet, if any, for the morphological left ventricle is a ventricular septal defect (Witham, 1957). Other anomalies may be associated and should be specified (Neufeld, Dushane, and Edwards, 1961a). DORV is distinguished from the tetralogy of Fallot with extreme dextroposition of the aorta, as there is loss of fibro-membranous continuity between the aortic and mitral valves and the aorta arises completely from the right ventricle with no override of the ventricular septal defect. Similarly, transposition of the great arteries with ventricular septal defect and extreme dextroposition of the pulmonary artery differs from DORV in which the pulmonary artery does not override the ventricular septal defect. Again, the absence of override is of particular importance in distinction of DORV from the Eisenmenger complex and the Taussig-Bing malformation (Van Praagh, 1968).

Other definitions for DORV have been used, reflecting the wide spectrum of bulboventricular malformations, and some are atypical cases or have features of other types of anomaly (Anderson *et al.*, 1974).

We describe in this paper some anatomical features in 27 cases of DORV, as defined above, which have particular relevance to the clinical diagnosis and surgical treatment of the condition.

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Material and methods

The 27 specimens of DORV were encountered among an anatomical review of 1610 hearts with congenital malformations: 910 from the Children's Hospital, Birmingham, and 700 from the Hospital Infantil, Madrid. Eleven of the patients died in the first 4 weeks of life and only 4 survived more than 1 year (Table 1).

All cases conforming to the definition given above (both great arteries arose entirely from the morphological right ventricle and showed no overriding) and having situs visceros-atrial solitus, laevocardia, and d-ventricular loop (Van Praagh *et al.*, 1964a) were included.

Particular attention was paid to the morphology of the right ventricle and outflow tracts, as described by Lev (1954) and by Van Praagh *et al.* (1964b). The relation of the great arteries to each other and to the conal anatomy were carefully noted, as was the relation of the semilunar valves to the atrioventricular valves. All associated anomalies were carefully examined.

Hypertrophy or dilatation of the cardiac chambers was judged subjectively. The left ventricle was considered to be slightly hypoplastic when the cardiac apex was formed by the right ventricle, and the anterior descending coronary artery passed to the left of the apex. A left ventricular cavity approximately 1 cm in diameter was assessed as moderately hypoplastic, and a minute cavity, 0.5 cm or less, was

TABLE 1 Associated anomalies

Case	Age	Venous anomaly	ASD	Mitral valve	VSD	Hypoplasia left ventricle	Pulmonary stenosis	Aortic stenosis	Persistent ductus arteriosus
<i>(a) Death in first month of life</i>									
64-355	1 d	—	+	Normal	+	Slight	—	+	+
70-94P	1 d	—	+	Hypo	+	Slight	—	+	+
66-72	2 d	LSVC	SA	Normal	+	Normal	+	—	+
67-83	2 d	TAPVD	+	Atresia	+	Moderate	—	+	+
69-129	2 d	LSVC	+	Atresia	+	Moderate	+	—	+
HRGG	3 d	—	—	Stradd	+	Slight	—	—	+
51-178	4 d	—	—	Hypo	—	Moderate	—	—	+
71-24	4 d	—	—	Atresia	—	Severe	—	+	+
70-422P	7 d	—	—	Atresia	+	Slight	—	+	+
66-297	11 d	TAPVD	SA	Atresia	+	Moderate	—	—	—
73-54P	16 d	—	—	Hypo	+	Moderate	—	+	+
<i>(b) Death between 1 month and 1 year of life</i>									
72-163P	1 mth	—	—	Atresia	+	Severe	+	—	—
AGG	1 mth	—	—	Stradd	+	Normal	—	+	+
70-625P	1 mth	—	+	Atresia	+	Severe	—	—	+
70-326P	1 mth	—	+	Atresia	—	Severe	—	—	—
69-132	2 mth	TAPVD	+	Hypo	+	Moderate	—	—	+
ML	2 mth	—	+	Atresia	+	Moderate	+	—	—
FFBJ	3 mth	—	+	Atresia	+	Severe	—	—	+
73-222P	3 mth	—	—	Atresia	—	Severe	+	—	+
MPFB	3 mth	—	CAVC	CAVC	CAVC	Moderate	+	—	—
63-368	5 mth	—	+	Atresia	—	Severe	+	—	+
66-48	5 mth	—	—	Stradd	+	Slight	+	—	—
70-85	8 mth	LSVC	—	Atresia	—	Severe	+	—	+
<i>(c) Death after 1 year</i>									
63-266	1	—	CAVC	CAVC	CAVC	Slight	+	—	—
67-246	3	—	—	Normal	+	Hypertrophy	—	+	+
71-99	4	PAPVD	+	Stradd	+	Slight	+	—	—
64-45	5	—	—	Normal	+	Normal	+	—	—

Abbreviations: LSVC, left superior vena cava; TAPVD, total anomalous pulmonary venous drainage; PAPVD, partial anomalous pulmonary venous drainage; ASD, atrial septal defect; SA, single atrium; CAVC, common atrioventricular canal; VSD, ventricular septal defect; PDA, persistent ductus arteriosus. +, defect present; —, defect absent; Stradd, straddling; Hypo, hypoplastic.

considered very hypoplastic. A great artery with a calibre one-third or less than the other great artery was considered to be hypoplastic. A semilunar valve with a circumference reduced by 50 per cent was considered stenotic.

In some cases, to reach a better understanding of the cardiac anatomy, the atria were separated from the ventricles and the ventricular musculature studied by blunt dissection following the planes and direction of the fibres, as described by Torrent Guasp (1966; F. Torrent Guasp, 1970, personal communication) and making serial records by drawings and photographs.

The relative position of the great arteries was noted *in situ* at the time of necropsy, and usually recorded photographically. In the few cases where this information was not available, the angio-

cardiographic records were used to orientate the specimen. The term normal relation of the great arteries was used when the aortic root was situated posteriorly and to the right of the pulmonary valve. We used d-lateral relation of the great arteries when both were situated at the same frontal level, the aorta being to the right. The term d-transposition was given to the cases with the aorta anterior and to the right of the pulmonary artery, regardless of the intracardiac conal anatomy.

The term crista supraventricularis was used for the curved muscular structure arching over the right ventricle. It runs from the bifurcation at the superior end of the septal band (trabecula septomarginalis) to gain the lateral wall of the ventricle. In the normal heart, it forms the posterior and inferior wall of the subpulmonary infundibulum.

FIG. 1 Double outlet right ventricle with double conus formed by a bifurcation of the crista supraventricularis. The conal septum is represented by the anterior branch of the crista. TV, tricuspid valve; M, mitral valve; CS, crista supraventricularis; Ao, aorta; PA, pulmonary artery (64.45 and 67.83).

FIG. 2 Subpulmonary conus. The aortic valve is in fibrous continuity with the tricuspid and the mitral valve straddles a ventricular septal defect (71.99).

FIG. 3 Subpulmonary conus. The crista does not originate from the interventricular septum but from the anterior ventricular wall (51.178).

FIG. 4 Subaortic conus. The mitral valve straddles the ventricular septal defect and is in fibrous continuity with the pulmonary valve (66.48 and HRGG).

Results

Morphology of right ventricle and infundibulum

(a) **DORV with double conus (complete subaortic and subpulmonary muscular infundibula)** There were 11 cases with this anomaly (Table 2). In the 7 with normally related great vessels or d-lateral transposition the crista supraventricularis, after leaving the interventricular septum, divided into two muscular branches which formed the aortic infundibulum. The aortic valve was separated from the tricuspid by the posterior branch, and the anterior branch separated it from the pulmonary valve. The posterior arch ended in the lateral wall of the ventricle, and the anterior arch ended in the right-anterior or anterior wall (Fig. 1). The pulmonary infundibulum was formed by the initial part of the crista, its anterior branch, the anterior ventricular wall, and the anterior part of the interventricular septum. Three of the 4 cases of aortic valve stenosis fell within this group (67-83 and 64-355 with normally placed arteries and 71-24 with d-lateral transposition). In the other 4 cases of double conus there was d-transposition, with the two branches of the crista forming a pulmonary conus. Three (70-85, MPFB, and 64-65) of the 5 cases with pulmonary valve stenosis had d-transposition. Five of the 11 cases of double conus had a ventricular septal defect between the tricuspid septal leaflet and the crista supraventricularis, and two cases had a common atrioventricular canal.

(b) **DORV with a subpulmonary conus** There were 10 such cases (Fig. 2). In 7 the aortic valve was in fibrous continuity with the tricuspid valve, in 2 others it was in continuity with the tricuspid and mitral valves, and in 1 case the aortic valve was in fibrous continuity only with the mitral valve across the ventricular septal defect. The pulmonary and aortic valves were separated from each other by the crista supraventricularis. In 8 cases, the crista formed an arch similar to that in a normal heart and the trabecula septomarginalis was well defined. In the other 2 the crista supraventricularis did not touch the interventricular septum. Its medial end originated not from the septum, but from the superior portion of the anterior wall of the right ventricle, just to the right of the interventricular septum (Fig. 3). It passed posteriorly and to the right, between the valves of both great arteries, and then ran laterally reaching the anterolateral wall of the right ventricle. There was a ventricular septal defect in all but one of this group; in 6 it lay between the crista supraventricularis and the posterior-superior branch of the septal band; in 2

it lay in the area of the endocardial cushions, being straddled by the mitral valve in 1 (Fig. 2); in 1 case the ventricular septal defect was placed between both branches of the septal band (70-422P). The relation of the great arteries was normal in 7 cases and was d-lateral in 2, both with pulmonary stenosis. Case 70-422P had a hypoplastic aorta located posterior and to the left of the pulmonary artery.

(c) DORV with a subaortic conus There were 4 cases, all with d-transposition of the great arteries. Two (Fig. 4) showed a ventricular septal defect anterior to the septal end of the crista, underlying the pulmonary outlet. The mitral valve straddled the defect, being in fibrous continuity with the pulmonary valve. The aortic infundibulum seemed to be formed by a division of the crista supraventricularis. In the other 2 cases the pulmonary artery was hypoplastic, its valve was in fibrous continuity with the tricuspid valve, and the crista supraventricularis arose not from the septum but from the anterior ventricular wall (Fig. 5).

(d) DORV with a common conus In 2 cases both great arteries arose from a common infundibulum, the pulmonary and aortic valves being separated from each other by a narrow fibrous ridge

FIG. 5 Subaortic conus. The crista is independent of the interventricular septum. Case 73-222P does not show a ventricular septal defect. Both have mitral atresia (72.163P and 73.222P).

(Fig. 6 and 7d). The common infundibulum resembled that of a normal right ventricle, being formed by the anterior-superior portion of the interventricular septum, the crista supraventricularis, and the anterior ventricular wall. The aortic valve and ascending aorta were hypoplastic in case 70-94P, and lay posteriorly and to the right of the pulmonary valve and artery. In Case ML, the pulmonary valve and artery were hypoplastic, lying posterior and to the left of the aorta. Both showed a ventricular septal defect between the tricuspid valve and the crista.

Relation between great arteries

Table 2 shows this relation, together with the

TABLE 2 Conal anatomy, valve stenosis, and relation of great arteries

Case	Continuity of valves	Double conus	Common conus	Aortic stenosis	Pulmonary stenosis	Relation of great arteries
64-355	—	+	—	+	—	Normal
70-94P	—	—	+	+	—	Normal
66-72	A-T-M	—	—	—	+	Normal
67-83	—	+	—	+	—	Normal
69-129	A-T	—	—	—	+	d-lateral
HRGG	P-M	—	—	—	—	d-transposition
51-178	A-T	—	—	—	—	Normal
71-24	—	+	—	+	—	d-lateral
70-422P	A-T	—	—	+	—	Inverted*
66-297	A-T	—	—	—	—	Normal
73-54P	A-T	—	—	+	—	Normal
72-163P	P-T	—	—	—	+	d-transposition
AGG	—	+	—	+	—	d-transposition
70-625P	A-T	—	—	—	—	Normal
70-326P	—	+	—	—	—	d-lateral
69-132	A-T	—	—	—	—	Normal
ML	—	—	+	—	+	d-transposition
FFBJ	—	+	—	—	—	d-lateral
73-222P	P-T	—	—	—	+	d-transposition
MPFB	—	+	—	—	+	d-transposition
63-368	—	+	—	—	+	d-lateral
66-48	P-M	—	—	—	+	d-transposition
70-85	—	+	—	—	+	d-transposition
63-266	—	+	—	—	+	d-lateral
67-246	A-M	—	—	+	—	Normal
71-99	A-T-M	—	—	—	+	d-lateral
64-45	—	+	—	—	+	d-transposition

*The aorta posterior and to the left of the pulmonary artery.

associated conal anatomy, and indicates whether there was semilunar valve stenosis or continuity between the atrioventricular and semilunar valves.

Out of 10 cases with a *normal relation* of the great arteries, 6 died in the first 2 weeks of life. Two had a double conus, 1 had a common conus, 1 had pulmonary stenosis, 5 had aortic stenosis, and 9 had ventricular septal defect. Among the 7 cases with *d-lateral relation* of the great arteries the age of

death was variable. Five had a double conus anomaly, 4 pulmonary stenosis, and 3 a ventricular septal defect. There were 9 cases with *d-transposition* and 8 died after the first month of life. One had a common conus, 4 a double conus, 7 pulmonary stenosis, 6 a ventricular septal defect, and 1 had preductal coarctation of the aorta. One patient (70-422P) died at 7 days; the aortic valve lay behind and to the left of the pulmonary (inverted position)

FIG. 6 Common conus. The crista (CS) and the trabecula septomarginalis, or septal band (TSM) are well defined (70.94P).

and there was mitral atresia with a ventricular septal defect, a displaced crista supraventricularis (not related to the septum), and hypoplasia of the aorta.

Associated anomalies

These are listed in Table 3 and tabulated in relation to age at death in Table 1.

A *ventricular septal defect* was found in 19 specimens. In cases 66–48 and HRGG it was located within the infundibulum of the pulmonary artery; the mitral valve overrode the ventricular septal defect and was in fibrous continuity with the pulmonary artery. In Case AGG, the ventricular septal defect was placed below the pulmonary infundibulum; here again the mitral valve straddled it, but it was not in continuity with the pulmonary valve. In 2 other specimens with an anterior ventricular septal defect (70–422P and 72–163P) the crista supraventricularis formed the infundibulum for a great artery but was not related to the interventricular septum, as described previously (see Fig. 5). In 10 cases the ventricular septal defect was located between the septal leaflet of the tricuspid valve and the crista supraventricularis. In another 2 cases (66–72 and 71–99) the ventricular septal defect lay in the area of the endocardial cushions. In both, the mitral and tricuspid valves were in fibrous continuity. Two of the cases (HRGG, AGG), with a ventricular septal defect overridden by the mitral valve, were included in a previous publication (Quero Jimenez *et al.*, 1973).

A *left superior vena cava* was present in 3 cases, all with pulmonary stenosis.

Anomalous pulmonary venous drainage was seen in 4 cases. The veins drained via the coronary sinus (2), directly into the right atrium (1), and the other showed partial drainage of the right pulmonary veins into the right atrium. All had a ventricular

FIG. 7 Four different patterns of conal anatomy. (a) Subpulmonary conus, the crista separates the semilunar valves. (b) The conal septum is represented by the anterior branch of the crista. (c) The crista does not originate from the septum but from the anterior ventricular wall, and separates the semilunar valves. (d) The crista lies behind the semilunar valves and there is no conal septum (69.132, 70.85, 70.422P, and ML).

septal defect, none had pulmonary stenosis, and the relation of semilunar valves was normal.

An *atrial septal defect* was encountered in 10 cases: ostium secundum in 9 and ostium primum in 1.

A *single atrium* was found in 2 specimens. One (66–297, Fig. 8) had a very large right atrium without any trace of a left atrium, left atrial appendage, or mitral valve. Both cases had anomalous pulmonary venous drainage, a ventricular septal defect, a subpulmonary conus, and normal relation of the great arteries. Both died during the first two weeks of life. The other 25 cases showed right atrial enlargement. There were single cases with absence of the coronary sinus (FFBJ), absence of the left atrial appendage (70–625P), and left juxtaposition of the atrial appendages (63–266). The foramen ovale was not identifiable in 2 specimens. The left atrium was normal in 8, hypertrophied in 2, and hypoplastic in 15 cases.

Mitral atresia was encountered in 13 cases; it was associated with an atrial septal defect in 7, a ventricular septal defect in 8, a double conus anomaly in 6, and pulmonary stenosis in 6. The *left ventricle* was underdeveloped in 24 cases. The 9 cases with a very hypoplastic left ventricle had mitral atresia without a ventricular septal defect.

TABLE 3 Associated anomalies

Anomaly	No. of cases
Left superior vena cava	3
Anomalous pulmonary venous drainage	4
Juxtaposition of atrial appendages	1
Single atrium	2
Atrial septal defect	10
Common atrioventricular canal	2
Mitral atresia	13
Straddling mitral valve	4
Ventricular septal defect	19
Pulmonary stenosis	12
Aortic stenosis	8
Persistent ductus arteriosus	18
Right aortic arch	1
Exomphalos with cardiac diverticulum	1

FIG. 8 Case 66-297. The left atrium is completely absent. Note how the anterior leaflet of the tricuspid valve attaches some chordae tendineae to the crista supraventricularis. RAPP= right atrial appendage.

Aortic valve stenosis was seen in 8 instances and was associated with infundibular stenosis (1 case), hypoplastic ascending aorta (3), hypoplastic aortic arch (2), and preductal coarctation (2). A *biventricular diverticulum*, which extended into an exomphalos, was present in Case 66-72 and was associated with a large ventricular septal defect, a single atrium, and a narrow tricuspid valve.

Discussion

Analysis of the findings in these 27 cases of DORV shows that none was free from serious associated malformations. This is perhaps not surprising in a necropsy study. Comparable data from successfully treated cases would be complementary and enable a

comprehensive study of the full range of this condition to be presented.

The commonest association was with ventricular septal defect (19 cases), and in DORV this constitutes the only outlet from the left ventricle. Eleven of these had significant defects of the atrial septum and in addition there were 2 cases of common atrioventricular canal. Nine had pulmonary stenosis, 7 had aortic stenosis, and 8 had mitral atresia. DORV with ventricular septal defect and no other significant anomaly is the most suitable combination for surgical correction, if this can be performed before the onset of irreversible hypertensive arteriopathic changes in the lung vessels, but there were no such examples in this series.

Pulmonary stenosis (12 cases) was the next com-

monest associated finding and was particularly frequent in the older children. It was observed in 8 of the 10 children of 3 months of age or older, but in only 4 of the 17 younger patients. Aortic stenosis, by contrast, was commoner in the younger group (7/17 and 1/10). In all but 1 of the 8 cases of aortic stenosis there was hypoplasia of the left ventricle and usually stenosis or atresia of the mitral valve.

A persistent ductus was seen in 18 cases. This was particularly common in the younger children as might be expected, not only because of the age effect, but because of the frequency of aortic stenosis in these babies. There was a persistent ductus arteriosus in 5 of the 10 children of 3 months of age or more, and in only 1 of the 4 who died after the age of 12 months.

Anomalous pulmonary venous drainage was found in 4 patients; 2 had mitral atresia and all had a ventricular septal defect. There were 3 patients with bilateral superior venae cavae; all 3 had pulmonary stenosis and 2 had mitral atresia.

These various findings underline the importance of the associated anomalies, and indicate that particular attention should be directed to the possibility of aortic, mitral, or pulmonary stenosis before surgical correction is undertaken. Anomalous pulmonary venous drainage should also be excluded. In addition, it would be desirable to assess the location of the ventricular septal defect.

The morphology of the infundibular region is of considerable practical importance in relation to diagnosis and to surgical treatment. The interpretation of the findings also has a bearing on our understanding of the normal development of the heart with particular reference to the crista supraventricularis and conal septum. The latter is the muscular tissue which normally separates the two semilunar valves and it normally contributes to the formation of the anterior part of the ventricular septum.

A complete muscular aortic infundibulum (Fig. 4 and 5) is usually associated with pulmonary stenosis, ventricular septal defect, and d-transposition (Table 4). The mitral valve, if it is not stenotic, tends to straddle the ventricular septal defect. The crista supraventricularis is dissociated from the anterior part of the ventricular septum and forms a conal septum by passing between the two semilunar valves (Fig. 5) or by giving rise to an anterior branch (Fig. 4).

A double conus (Fig. 1) is commonly associated with either pulmonary or aortic stenosis (Table 2). The aorta usually is in either the d-lateral or the d-transposition situation (Table 4) and the posterior valve tends to be stenosed. The crista supraventricularis forms the posterior wall of the conus and its anterior branch forms a conal septum.

TABLE 4 *Relation of conal anatomy and semilunar valve stenosis to position of great arteries*

	Normal d-lateral		d-transposition	Total
Subaortic conus	0	0	4	4
Subpulmonary conus	7+1*	2	0	10
Double conus	2	5	4	11
Common conus	1	0	1	2
Aortic stenosis	5+1*	1	1	8
Pulmonary stenosis	1	4	7	12
No stenosis	4	2	1	7
Total	10+1*	7	9	

*1 case with inversion of aorta.

tricularis forms the posterior wall of the conus and its anterior branch forms a conal septum.

A common conus, enclosing the origin of both major vessels with no intervening muscular elements, was seen only twice (Table 2, Fig. 6 and 7d). There was no conal septum between the two valves, but the crista supraventricularis was well developed and formed the posterior wall of the conus.

In the 10 cases of DORV with a subpulmonary conus (Fig. 2, 3, and 7c) the crista formed a conal septum between the two semilunar valves. It usually formed an arch of normal appearance with a well-defined trabecular septomarginalis and a ventricular septal defect was common. In 2 cases (Fig. 3 and 7c) the crista did not fuse with the ventricular septum.

The variations in the infundibular morphology in DORV suggest that the crista develops normally, not from the left anterior conal ridge as commonly understood, but from the bulbus cordis. The four main patterns observed are shown in Fig. 7. They show that the crista is always represented even though the conal septum may be absent. This independence may indicate different origins—the conal septum from the conal ridges and the crista from the bulbus.

The pattern seen in our 2 cases with a common conus appears to be related to that in common truncus arteriosus. Case 70-94P (Fig. 6) was very similar to that described by Van Praagh and Van Praagh (1965) as common truncus with persistence of the VIth arch. It showed complete truncal septation, but Van Praagh's lacked septation in its lowest portion. Case ML (Fig. 7d) showed a similar relation to common truncus with persistence of the IVth arch, and fibrous discontinuity between the truncal and the atrioventricular valves.

The conal anatomy is related to the position and calibre of the great arteries. Table 4 shows that the anterior vessel always has a muscular conus, though

TABLE 5 *Relation of conal anatomy and semilunar valve stenosis*

Conus	Stenosis			Total
	Nil	Pulmonary	Aortic	
Subaortic	1	3	0	4
Subpulmonary	4	3	3	10
Double	2	5	4	11
Common	0	1	1	2
Total	7	12	8	

in some cases this is part of a common or double conus. Looked at another way, it is clear that normal position of the great vessels is usually associated with a pulmonary conus, but d-lateral, and to a lesser degree d-transposition, tend to be associated with a double conus. Table 5 shows that a stenotic outlet is usually related to the posterior vessel. In other words, normal relation of the two arteries usually implies a single pulmonary conus with or without aortic stenosis, and in d-transposition there is usually a double or single aortic conus with pulmonary stenosis.

Our 2 cases with a common atrioventricular canal both had a double conus with pulmonary stenosis but this cannot be said to establish a consistent pattern.

From the clinical point of view, 12 of our cases had features of Fallot's tetralogy and 7 showed the picture of a large ventricular septal defect, both being well recognized modes of presentation in DORV (Neufeld *et al.*, 1961a, b). More than a third of our cases, however, died within the first 2 weeks of life, with the predominant clinical features of hypoplastic left ventricle and aortic stenosis.

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