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The nomenclature of duct-dependent lesions, broadening the categories for a better clinical practice



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ABSTRACT

Ductus arteriosus-dependent lesion includes two well-established categories, the right sided obstructive lesions such as pulmonary atresia (duct-dependent pulmonary circulation) and the left sided obstructive (duct-dependent systemic circulation) lesions ranging from hypoplastic left heart syndrome to severe preductal coarctation. Dextro-transposition of the great arteries (D-TGA) lies in a gray area, as it is considered by some old classifications as a duct-dependent pulmonary circulation lesion and by others as a new category of duct-dependent mixing lesion. We suggest adding a new functional classification, where the category of duct dependent systemic circulation/perfusion remains unchanged, and whereby duct-dependent pulmonary lesions are renamed into a broader category of duct dependent oxygenation with two sub-categories: mixing defects such as D-TGA, and defective oxygenation due to right sided obstructive lesions such as pulmonary atresia. We believe that this re-classification will cover the anatomical aspects and clinical implications of these lesions and will facilitate their management by the residents which are the primary encounter physicians in many of the developing countries.

Keywords: Nomenclature of duct dependent lesions; Duct-dependent systemic perfusion and oxygenation; D-TGA.

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BACKGROUND

Congenital heart defects (CHDs) represent a spectrum of structural abnormalities affecting the heart's development, ranging from minor defects to complex, life-threatening conditions. Critical CHD are lesions presenting shortly after birth and accounting for 50% of infant mortality due to congenital defects. The most important constituent of this category is the duct-dependent lesions. The important hemodynamic change which occurs after birth is the transition from fetal circulation to postnatal circulation and during this transition, newborns with CHD's are vulnerable thereby increasing their dependence on central shunts mainly patent ductus arteriosus (PDA) to ensure ample blood flow between the systemic and pulmonary circulation. These lesions are collectively known as "Ductal-dependent lesions".¹

In a normal heart, PDA typically closes shortly after birth as a part of normal physiological changes due to

the transition from fetal to neonatal circulation. However, since right or left obstructive lesions heavily rely on the blood flow through the PDA for adequate circulation between the systems, patients present with severe cyanosis, shock or collapse. Hence, duct-dependent lesions (DDL) stand out among critical CHD's as it requires immediate clinical attention and intervention.²

MAIN BODY

The current classification: an anatomic classification

Lesions where the systemic circulation is interrupted by the presence of severe left sided obstruction at the level of the left ventricle as in critical aortic stenosis or hypoplastic left heart syndrome or at the level the aorta as in interrupted arch or preductal aortic coarctation, ductal flow from the pulmonary artery ensures the maintenance of the distal systemic blood flow. Disruption of this delicate balance can result in compromised perfusion

of vital organs (systemic perfusion). As the PDA constricts or closes after birth, these conditions may lead to systemic hypotension, shock, or collapse.³

Inversely, defects with varying degrees of right-sided heart obstruction as pulmonary atresia, critical pulmonary valve stenosis or pulmonary atresia with intact ventricular septum are commonly called Duct Dependent Pulmonary lesions. Such defects mainly rely on duct dependent pulmonary flow in which pulmonary blood flow or oxygenation is dependent on the flow arising from the aorta to the pulmonary artery via the ductus. Typically, these conditions lead to poor oxygenation of blood causing cyanosis as the PDA closes after birth.⁴

D-TGA wandering between different classifications

Cyanosis, also results from another duct-dependent lesion which is Dextro-Transposition of the great arteries (D-TGA), where the ventriculoarterial connections are "misplaced" or

Table 1. Current classification of Duct dependent lesions

Category	Category 1 Duct dependent systemic perfusion or circulation	Category 2: Duct dependent pulmonary circulation	Category 3: Duct-dependent mixing
Included CHD	Left sided obstructive lesions e.g.: Hypoplastic left heart syndrome Interrupted aortic arch	Right sided obstructive lesions: Pulmonary atresia, critical pulmonary stenosis	D-TGA

Table 2. Suggested classification for duct-dependent lesions

Category	Category 1: Duct-dependent systemic perfusion	Category 2: Duct dependent oxygenation:	
Included CHD	Left sided obstructive lesions e.g.: Hypoplastic left heart syndrome Interrupted aortic arch	Sub-category A: Right sided obstructive lesions Pulmonary atresia Critical pulmonary stenosis	Sub-category B: Mixing lesion D-TGA

“transposed” across the interventricular septum. As a result, the pulmonary artery arises from the morphologic left ventricle and the aorta from the morphologic right ventricle. In D-TGA the cause of cyanosis and duct dependence is the presence of two parallel non communicating loops, a fully deoxygenated right-sided loop and left-sided oxygenated loop, the right loop carries desaturated blood to the body via the aorta. Blood mixing between these two circulations, which takes place via PDA, atrial septal defect (ASD) or Patent foramen ovale (PFO), or VSD, is essential for survival in neonates with TGA. There is conflicting literature on the classification of D-TGA, some of the relatively old literature tended to classify it as a duct-dependent pulmonary circulation, some others as an independent entity, in the presence of a significant PFO or VSD, it is being classified as a duct independent mixing lesion alongside total anomalous pulmonary venous return and truncus arteriosus or in the absence of the latter as duct-dependent mixing lesions. (Table 1 shows the current classification).⁵

Our suggested classification

Despite being currently classified as an independent category, we believe that D-TGA has the same clinical spectrum as duct dependent pulmonary circulation lesions (right sided obstructive lesions), they both present with desaturation, as a result of defective oxygenation, and they both share the fact that the maintenance of the ductus improves their oxygenation. And despite the discrepancy in the

mechanism by which the ductus in each of those lesions, maintains, the oxygenation, we believe that including them in a single functional category is essential for the pediatric residents and trainees to draw their attention, to the same pattern of early cyanosis after birth, which differs from the early shock and hypoperfusion presented by duct dependent systemic lesions. Thus our suggestion (as presented in table 2, is to return to a two category model of duct dependent lesions, where duct dependent systemic lesions remains unchanged, and duct dependent pulmonary lesions, gets renamed into duct dependent oxygenation lesions, which will include two sub-categories: right sided obstructive lesions (as pulmonary atresia) and defective mixing (D-TGA).⁵

CONCLUSION

In conclusion, the transition from “duct-dependent pulmonary and systemic lesions” to “duct-dependent oxygenation and systemic perfusion” signifies a progressive step in medical terminology. By understanding a more comprehensive and physiological perspective of congenital heart defects. This change in nomenclature recognizes the broader meaning which facilitates a more integrated approach to diagnosis, management, and treatment options. Moreover, enhancing communication between healthcare professionals and patients. Using terms such as “duct-dependent oxygenation and systemic perfusion” could promote clearer explanations to patients and their

families, developing a more informed and engaged healthcare experience. The current terminology limits the broader implications of duct-dependent lesions. This shift reflects a deeper understanding for cardiovascular physiology and the critical role of the ductus arteriosus in sustaining adequate oxygenation and perfusion.

CONFLICTS OF INTEREST

The authors declare no conflict of interest.

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I wanted, as a first author, to dedicate my work to compassionate people, those who feel the pain of others and their struggles. We need a world with more compassion, and less judgement.

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