the Child Dental Patient with

Shone's Syndrome

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Shone's syndrome, named after the doctor who first described it in 1963, is a rare association of multiple obstructions at the entrance and exit of the left ventricle. It is also known as *Shone's complex* as it is related to four different abnormalities:

1. aortic coarctation;
2. (membranous or muscular) subaortic stenosis;
3. supravalvular mitral membrane;
4. parachute mitral valve;

1. DOI: 10.1016/j.radcr.2018.10.004 Alan True et al., Computed tomography imaging characteristics of shone syndrome, Radiology Case Reports, Volume 14, Issue 2, 2019,
2. DOI: 10.1016/0002-9149(63)90098-5 John D. Shone, et al., The developmental complex of "parachute mitral valve," supravalvular ring of left atrium, subaortic stenosis, and coarctation of aorta VOLUME 11, ISSUE 6, P714-725, JUNE 01, 1963

Since its description in the 1960s, Shone's complex definition has been expanded to include bicuspid aortic valve with aortic stenosis and VSD (ventricular septal defect).

Symptomatology depends on the severity of the obstruction.

This syndrome is characterized by left ventricular filling difficulties during diastole due to mitral insufficiency, and left outflow difficulties during systole due to subaortic stenosis.

It is most frequently diagnosed in its incomplete form.

The prognosis is **unfavorable**.

1. Lee A Fleisher, Anesthesia and Uncommon Diseases Elsevier Health Sciences, 20 apr 2012
2. Jing Ping Sun et al, Comparative Cardiac Imaging: A Case-based Guide John Wiley & Sons, 20 feb 2018

Aortic Coarctation (AOC): congenital cardiac anomaly consisting of localized narrowing of the aortic lumen causing hypertension of the upper limbs, left ventricular hypertrophy, and hypoperfusion of the abdominal organs and lower limbs.

DOI: 10.5830/CVJA-2017-053 CardioVascular Journal of Africa, Volume 29, No 4, July/August 2018

Subaortic stenosis: a rare cardiac malformation that can manifest as a muscular ridge or thin membrane and causes the obstruction of the left ventricular outflow tract below the aortic valve.

1. Catherine M. Otto Ecocardiografia clinica: Sesta edizione Edra, 2018

Mitral Supravalvular Membrane: anomaly characterized by an abnormal ridge of connective tissue on the atrial side of the mitral valve. The result is a circular ring that can encroach on the orifice of the mitral valve and adhere to the mitral valve leaflet, thus restricting its movement. It usually causes significant obstruction of the flow from the left atrium to the left ventricle.

1. M Satpathy, Clinical Diagnosis of Congenital Heart Disease JP Medical Ltd, 30 ago 2015

Parachute mitral valve: an anomaly of the mitral valve in which all the tendinous chords of the valve, which can be shorter and thicker, fit into a single, abnormal papillary muscle. This is usually the cause of mitral stenosis.

1. Michael A. Gatzoulis et al, Cardiopatie congenite dell'adulto: Una guida pratica Springer Science & Business Media, 29 apr 2007

Shone's Syndrome & Oral Health

Children with congenital heart defects (CHD) are exposed to many oral health risk factors:

- *Chronic intake of sugary medicines*;
- *A greater tooth enamel defect susceptibility.* The systemic effect of CHD has been assumed to induce enamel defects during the enamel formation period as ameloblasts are highly sensitive to metabolic changes;
- *Lack of attention to dental care and oral hygiene* due to increased concern about heart disease.

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Data on parents' awareness of the importance of proper oral hygiene and preventive dentistry show that parents receive little **or no** information on this subject.

1. DOI: 10.1007 / s00784-017-2256-2 Koerdt, S., Hartz, J., Hollatz, S. *et al.* Dental prevention and disease awareness in children with congenital heart disease. *Clin Oral Invest* **22,** 1487–1493 (2018)
2. DOI: 10.17796 / jcpd.33.4.2j108w0225241867 Kavita Rai and Supriya S and Amitha Hegde, Oral Health Status of Children with Congenital Heart Disease and the Awareness, Attitude and Knowledge of their Parents, Journal of Clinical Pediatric Dentistry, 2009

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