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Hypoplastic Right Heart Syndrome is a Rare Disorder – Case Report

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INTRODUCTION

Hypoplastic Right Heart Syndrome (HRHS) is a condition that is even more rare than Hypoplastic Left Heart Syndrome (HLHS). HRHS refers to the underdevelopment of the right side structures of the heart, which means that the chambers, valves and related blood vessels on the right side of the heart are malformed. This malformation involves the pulmonary valve atresia which has not formed, a very small right ventricle, a small tricuspid valve and a small hypoplastic pulmonary artery. As the ventricle has failed to grow and develop the ventricles muscle structure is poor, so additional problems are encountered as the heart attempts to pump blood to the pulmonary valve for transfer to the lungs. The proper amount of blood pumped from the right atrium is not sufficient and this causes the blood to be not pumped efficiently to the lungs.

CASE REPORT

A newly delivered newborn baby developed dyspnoea,bluish discolouration of lips, tongue ,desaturation. on examination child is dyspnoeic,cvs –s1s2 +,short systolic murmur in left 2nd intercostal space rs – vbs ,no added sounds, spo2 80 %
Abdomen –soft, no organomegaly.
ECG –sr,hr 158 /m, p pulmonale,no st –t changes, chest x ray –pulmonary olegemia, echo – situs solitus, rv is very small , pulmonary atresia, small pulmonary artery small pfo, pda with left to right shunt , ias thinned, aneurysmal, no asd,no vsd normal lv function.
HB 16.7 gm%,wbc 24700,platelet count 255000,crp 1mg/l,
Child is diagnosed to have hypoplastic right heart syndrome.
Child managed with iv antibiocts, o2, other supportive medications and child is being reffered to paediatric cardiologist / higher centre for further management .

CONCLUSION

Hypoplastic Right Heart Syndrome is a Rare Disorder.

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