Double outlet right ventricle

How does a normal heart work?
A normal heart has two smaller upper chambers called the atria and two larger lower chambers called ventricles. It has four valves, one at the entrance to each chamber. They stop the blood flowing backwards and keep the blood moving forwards through the heart. The left side of the heart receives oxygen-rich blood from the pulmonary veins. The heart then pumps it through the aorta and around the body. The right side of the heart receives blood from the body that has used up its oxygen, so the blood is pumped to the lungs through the pulmonary artery to replenish its supply. Blood is then pumped to the heart’s left side, where the cycle begins again.

What is double outlet right ventricle?
In double outlet right ventricle, the body’s two biggest arteries (the aorta and the pulmonary artery) originate in the right ventricle. Blood from the left ventricle passes through a small opening in the wall between the left and right sides of the heart into the right ventricle to reach the two arteries. This abnormal opening is known as a ventricular septal defect (VSD). Blood flow from the heart to the lungs often increases and is under very high pressure.

Are there any symptoms?
The symptoms of double outlet right ventricle develop in the first weeks of a baby’s life. Your child may suffer blueness of the skin (cyanosis) and breathlessness, and he or she may be unable to put on weight. The symptoms will vary depending on the extent of the abnormality or the presence of other defects.

How is it treated?
Your child can have surgery to repair the defect. The age at which this will happen will depend on the child’s individual condition. In most cases, a complete repair is possible, although your child may have to have several operations to achieve this. Your child may then be well enough to go to school and lead a near-normal life as he or she grows up.

In a small number of cases, a complete repair is not possible and the aim will be to make your child’s heart work as efficiently as possible. Several staged operations may be needed, one of which could be the ‘Fontan’s procedure’. After this operation, ‘blue blood’ (low in oxygen) can flow directly to the lungs bypassing the heart. The ventricles then pump oxygen-rich blood (red) round the body. While this operation cannot correct the original problem, it may reduce the blue appearance of your child’s skin.
**What are the associated problems?**
Many babies affected by this condition may also suffer pulmonary stenosis or valve abnormalities. In these cases, your baby may need to have surgery in the first few weeks of their life.

**Further information:**

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