Hemivertebra

Patient Information Series – What you should know, what you should ask.

What is hemivertebra?

Hemivertebra is a congenital (present from birth) anomaly of the spine in which only one half of the vertebral body develops. It occurs in about 3 in 10,000 births. It is a common cause of congenital scoliosis where the spine is curved sideways. Children with congenital scoliosis sometimes have other health issues, such as kidney or bladder problems.

How does hemivertebra happen?

Our spine is usually made up of 33 vertebrae stacked in a column. At about 6 weeks' gestational age, each vertebra has two areas at its sides where the bone develops and hardens, called lateral ossification centers. A hemivertebra results from the failure of one of these lateral centers to develop, so the vertebra is missing one side. The defective vertebra impacts on the configuration of the spinal column by acting as a wedge in the spine, leading to excessive lateral curvature (scoliosis), or too great a curve away from the side on which it is present.

Should I have more tests done?

Tests to ask about include an amniocentesis to look for problems with the number of chromosomes and some underlying genetic conditions. Amniocentesis is a test where a needle is used to take some of the fluid surrounding the fetus in the womb. Other genetic tests may be offered, such as Chromosomal Microarray (CMA, or "chip"), or exome sequencing, which look more closely at the genetic make-up of the fetus, if this testing is available.

Because Hemivertebra has been described in association with various other anomalies (such as other abnormalities of the bones and muscles, including those of the spine, ribs, and limbs), you should also ask for a detailed ultrasound examination. Use of 3D imaging can also help in determining the presence of hemivertebra and the resulting scoliosis. This can be used to have a prenatal consultation with a paediatric orthopaedic surgeon to understand the extent of the condition and postnatal management. Hemivertebra is also associated with congenital

syndromes including Jarcho-Levin, Klippel-Fiel, Aicardi syndrome, and VACTERL association, so genetic counselling could be of benefit to you.

What does it mean for my baby after it is born?

Congenital scoliosis resulting from hemivertebra is not painful and the outcome depends on further progression during development and growth of the baby.

Left untreated, 25% of patients with congenital scoliosis show no progression, 50% progress slowly, and 25% progress rapidly during growth. The treatment depends upon how likely the spinal curve is to worsen and can vary from nonsurgical to surgical options. Timely follow up with the orthopaedic surgeon is suggested. In mild cases, observation can suffice. However, in severe cases the treatment approach is usually surgical in nature. Spinal fusion is considered the treatment of choice, in which the abnormally growing vertebra are fused together to promote healing of single solid bone. This is usually performed before significant deformity occurs and prevents further worsening of the spinal curvature.

Will it happen again?

This is uncertain and depends on whether a genetic cause was determined. There may be an increased risk of neural tube defects in siblings, but the risk would be low. In subsequent pregnancies, a targeted study can be done right from the first trimester to detect the presence of hemivertebra and the consequent scoliosis.

What other questions should I ask?

- Is the Hemivertebra isolated or associated with other anomalies?
- Should I do a genetic test?
- How often will I have ultrasound examinations done?
- Is surgery after pregnancy available?
- Where should I deliver?
- Where will the baby receive the best care after it is born?
- Can I meet in advance the team of doctors that will be looking after my baby when it is born?

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