Renal agenesis (missing kidneys)

What are kidneys?
Most of us have two kidneys which are usually located either side and in front of our lumbar spine. A normal kidney contains about a million nephrons which are pipes which filter the waste products from our blood and control the amount of water in our bodies.

What is Renal Agenesis?
For reasons which are not clear at this time the process may go wrong and either one or both kidneys may fail to develop. In addition occasionally kidneys may be in the wrong place (ectopic kidney). About one in every 300 children is born with a single kidney (Unilateral renal agenesis). As we only need about half of one normally functioning kidney to lead a normal healthy life having a single kidney is of little significance unless at some stage in the future that kidney is damaged. Unfortunately in one in every 3000 pregnancies neither kidney forms. (Bilateral renal agenesis which is known as Potter’s syndrome).

How is renal agenesis detected?
The first clue that there might be a problem is when a patient undergoes an antenatal scan and the sonographer finds no fluid (liquor) around the baby. Up until 15 -16 weeks gestation the majority of fluid comes from simple diffusion of fluid through the baby’s skin or the placenta. The baby will start to produce urine and empty its bladder into the amniotic fluid from about 11 weeks. By 16 weeks the majority of the amniotic fluid is from the baby’s bladder, therefore if there are no kidneys and therefore no urine production there will be no fluid around the baby. Normal amounts of fluid are needed to allow us to get good view of the baby. Without it we have problems checking all of the structures we usually look at including the kidneys.

Are there other causes of reduced fluid around the baby?
There are other causes of reduced or absent fluid around the baby. These include-
- A hole in the membranes (the sac) around the baby. In this case any fluid produced simply leaks out vaginally. Although most women are aware of this loss this is not always the case.
- Two abnormal kidneys or an inability of the baby to be able to pass urine because of a blockage between the bladder and the outside.
- Reduced fluid can be caused by severe growth restriction.

How do we differentiate between renal agenesis and other causes of reduced fluid?
In renal agenesis we should not be able to see a bladder (not because it is not there but because it contains no fluid which is how we usually recognise a bladder). However, it should be recognised that the diagnosis can sometimes be very difficult to make.
What is the effect of having no fluid around the baby?
The amniotic fluid serves a number of purposes. It provides a cushioned environment in which the baby can grow and move freely. If there is no fluid then the baby’s limbs are often held in a fixed position which due to the lack of movement possible results in conditions such as talipes. This is where the foot is turned over at the ankle. This is a common condition seen in other circumstance and is correctable. No fluid also results in the baby’s face being “moulded” and these babies usually have a small nose and jaw and low set ears.
The biggest problem however, is with the lungs. Normally babies breathe in and out amniotic fluid. This is essential for normal lung development. If this does not occur than the baby develops a condition called pulmonary hypoplasia, which is just a medical term for small lungs. The lungs of a baby with no kidneys are so small at birth that even if we give the baby 100% oxygen to breathe it cannot take in enough oxygen to survive and therefore these children die not because of kidney failure but because of their small lungs.

Can we offer any treatment for this?
In theory doing the reverse of an amniocentesis and putting fluid into the sac could prevent this occurring. This has not been demonstrated to work in practise and would carry a risk of pregnancy loss every time it was undertaken. Although it can be done the fluid only last a matter of a couple of days and therefore the procedure has to be repeated frequently. Even if it were successful then it does not cure the underlying problem of no kidney which would mean dialysis from birth and renal transplantation which is not very successful in children under one year old.

What happens if we are unsure of the diagnosis?
As the ultrasound views are poor we might be unsure about the diagnosis of renal agenesis. However, we can be sure about the lack of fluid and it is this feature as much as anything which will determine the outcome. If there is a hole in the membrane we would expect to clearly see the kidneys and if we were struggling to see these then at least we would expect to see the bladder fill and empty. There are other tests we might consider. One of which is amnioinfusion. This is the procedure whereby we put fluid back into the sac. This has the advantage of giving us much better pictures. In addition if there is some functioning renal tissue somewhere in the baby we would expect over 20-30 minutes to see the bladder fill and empty.

What are my options?
If we have made a diagnosis of renal agenesis then you will be offered the opportunity of either continuing with the pregnancy or opting for a termination. If you continue one often finds that delivery occurs a little early, usually at around 35 weeks. We would suggest that we should only listen in occasionally to be baby’s heart beat during labour as we would not want to perform a caesarean section as this would not improve the baby’s outcome. We would arrange to have a baby doctor present at the time of delivery. Your baby may be born alive but if the lungs are very small then we would not expect the
baby to live for more than a short period of time. There is no reason why you should not be able to cuddle your baby at this time. The other option is obviously to choose a termination of pregnancy.

**Will it happen again?**
Renal agenesis can occur for many different reasons. As a general rule it is unlikely to happen again but we may well to do further tests including scanning both parents kidneys before giving a more precise risk of recurrence risks.

**When could it be detected?**
It is possible to see kidneys from as early as 12-14 weeks in the pregnancy. At 12 weeks we would usually be able to see a fetal bladder. So in a future pregnancy one would hope to be able to be reassuring from as early as 12-14 weeks that all was well.