CONDITION

Potter syndrome

Potter syndrome is a pattern of fetal development caused by too little amniotic fluid, often because the kidneys did not form.

WHAT IT IS
Developmental effects of severe, prolonged low amniotic fluid

BODY SYSTEM
Kidneys, urinary system, lungs, face, and limbs

MAIN SYMPTOMS
Underdeveloped lungs, breathing failure, limb differences, facial features

WHAT CAUSES IT
Kidney abnormalities, urinary blockage, or early membrane rupture

HOW IT IS DIAGNOSED
Prenatal ultrasound and examination after birth

MAIN TREATMENT
Specialist pregnancy care, breathing support, kidney care, or comfort care

CAN IT BE CURED?
No - established organ changes cannot be reversed

IS IT AN EMERGENCY?
Yes - severe breathing difficulty after birth

LONG-TERM OUTLOOK
Often fatal before or shortly after birth

Overview

Potter syndrome is a pattern of physical and medical problems caused by having very little or no amniotic fluid for a prolonged time during pregnancy. Amniotic fluid is the liquid surrounding a fetus in the uterus.

Clinicians may describe this as the Potter sequence because one problem leads to several others. Too little fluid limits the space available for movement and, more importantly, interferes with lung development.

After the first part of pregnancy, most amniotic fluid comes from fetal urine. The fetus swallows the fluid, and the kidneys and bladder help maintain its circulation. If both kidneys are absent or do not work, little or no urine enters the amniotic sac. Severe urinary tract blockage or an early leak of amniotic fluid can cause a similar pattern.

The most serious effect is pulmonary hypoplasia , meaning the lungs are much smaller and less developed than expected. Severe pulmonary hypoplasia can make breathing impossible or extremely difficult after birth. Potter syndrome is therefore often fatal before birth or during the newborn period.

The term was originally linked closely with bilateral renal agenesis, in which both kidneys are absent. It is now also used for the same pattern when severe, long-lasting fluid loss has another cause.

Symptoms

Potter syndrome develops before birth. The first sign is often very low amniotic fluid found during an ultrasound rather than something the pregnant person can feel.

Ultrasound or examination may show:

Limited space in the uterus can affect the face and limbs. Possible features include a flattened nose, a small or receding chin, low-set ears, folds near the inner corners of the eyes, clubfeet, or joints held in fixed positions. These features do not by themselves confirm Potter syndrome.

After birth, the main concern is breathing. A newborn may breathe rapidly, struggle to move air, have a bluish or grey skin colour, or be unable to maintain oxygen levels. If the kidneys are absent or severely impaired, the baby may make little or no urine.

The findings are not identical in every pregnancy. Their severity is influenced by how early the fluid became low, how long it remained low, the underlying cause, and how much lung and kidney function developed.

- Very little or no amniotic fluid
- Missing, abnormal, or enlarged kidneys
- A bladder that does not fill, or an enlarged bladder caused by blockage
- Underdeveloped lungs
- Reduced fetal movement or unusual positioning of the limbs

Causes and risk factors

The most recognized cause is bilateral renal agenesis , meaning neither kidney formed. Without functioning kidneys, the fetus cannot make the urine needed to maintain amniotic fluid later in pregnancy.

Other causes include:

Some kidney and urinary tract abnormalities occur on their own. Others are associated with a chromosome difference, a genetic condition, or abnormalities in several organ systems. Potter syndrome itself is not always inherited.

The likelihood of the same problem happening in another pregnancy depends on its cause. For example, the recurrence risk may be different for an isolated kidney-development problem than for an inherited kidney disorder. A genetic counsellor can review the pregnancy history, test results, and family history to give a more specific assessment.

Nothing a pregnant person did during ordinary daily life causes the characteristic Potter pattern from missing kidneys. When early membrane rupture is responsible, a clear reason for the rupture may never be found.

- Severe malformation or disease affecting both kidneys
- A blockage that prevents urine from leaving the bladder
- Rupture of the amniotic sac early in pregnancy
- Less commonly, severe problems affecting placental blood flow

Diagnosis

Potter syndrome is usually suspected during a prenatal ultrasound. The scan measures amniotic fluid and looks at the kidneys, bladder, lungs, urinary tract, growth, and other organs.

Repeated ultrasound examinations may be needed. Very low fluid can make images harder to obtain because fluid normally creates space around the fetus. In selected cases, fetal magnetic resonance imaging, or MRI, may provide more information about the lungs or anatomy.

Doctors will try to identify why the fluid is low. Important findings include whether both kidneys can be seen, whether the bladder fills over time, and whether there is a urinary blockage. They will also consider whether amniotic fluid is leaking through the vagina.

Chromosome testing or broader genetic testing may be offered when ultrasound shows kidney abnormalities, additional birth differences, or a possible inherited condition. These tests may explain the cause, but a normal result does not rule out Potter syndrome.

After birth, diagnosis is based on the baby's breathing, physical examination, urine production, and imaging of the kidneys and urinary tract. Blood tests can assess kidney function. If the pregnancy ends in fetal loss or the baby dies, examination of the placenta and, when the family agrees, a post-mortem examination may provide information that imaging could not.

Treatment and management

No treatment can replace kidneys that did not form or reverse lungs that are already severely underdeveloped. Care therefore focuses on understanding the cause, preparing for birth, and making decisions that reflect the expected medical outcome and the family's values.

During pregnancy, care commonly involves maternal-fetal medicine, fetal imaging, neonatology, kidney specialists, genetic services, and supportive or palliative care teams. Follow-up ultrasound can clarify fetal growth, fluid levels, anatomy, and likely lung development. The team may also discuss where and how birth should occur.

Procedures that add fluid to the amniotic sac have been studied, including repeated amnioinfusions . Research has shown that this may allow lung development and short-term survival in some carefully selected pregnancies involving bilateral renal agenesis. However, it does not create kidney function. Surviving babies require intensive care and dialysis, and the procedure carries substantial burdens and pregnancy risks. It remains specialized care rather than a cure or routine treatment.

If there is a treatable fetal urinary blockage, a fetal medicine centre may assess whether an intervention could help in that specific situation. Not every blockage is suitable for treatment, and restoring urine flow may not restore kidney function that has already been lost.

After birth, options may include:

Comfort care treats pain, breathlessness, agitation, and family distress. It can include holding, feeding when safe, warmth, memory-making, and spiritual or cultural support. Choosing comfort-focused care is active medical care, not abandonment.

- Intensive breathing support
- Treatment of low blood pressure and other newborn complications
- Dialysis when kidney failure is present and lung function allows survival
- Later kidney transplantation for a small number of children who become eligible
- Comfort-focused care when life-prolonging treatment cannot provide a realistic benefit

Complications

Severely underdeveloped lungs are the main life-limiting complication. Even a breathing machine cannot always move enough oxygen through lungs that have too little functioning tissue.

Other possible complications include:

Families may also experience grief, anxiety, depression, or trauma following the diagnosis, pregnancy loss, or newborn death. These effects deserve care in their own right. Bereavement services, counselling, peer support, and follow-up with the pregnancy team may help.

- Fetal loss or death shortly after birth
- Kidney failure with little or no urine production
- Air leaks from fragile lungs during breathing support
- Joint stiffness, clubfeet, or other positioning differences
- Problems related to premature birth
- Complications from prolonged intensive care or dialysis

Prevention

Potter syndrome usually cannot be prevented. In particular, there is no known action that can make absent kidneys develop later in pregnancy.

Routine prenatal care may identify low amniotic fluid or fetal abnormalities, but early detection is not the same as prevention. If fluid is leaking, timely assessment may help manage infection, premature labour, and other pregnancy risks, although it cannot always prevent the Potter pattern.

When a genetic or chromosome cause is found, genetic counselling can explain reproductive options for future pregnancies. In a later pregnancy, early targeted ultrasound or diagnostic testing may be offered based on the specific cause. These steps can provide earlier information but do not guarantee prevention.

Living with it and what to expect

Potter syndrome is most often diagnosed during pregnancy, so families may have to make major decisions while still processing difficult news. The medical team may discuss continuing the pregnancy, pregnancy termination where legally available, plans for intensive newborn treatment, and perinatal comfort care.

The outlook is especially poor when both kidneys are absent and there is prolonged absence of amniotic fluid early in pregnancy. Some fetuses die before birth, and many affected newborns die because their lungs cannot support breathing.

Survival is possible in selected situations, particularly when some lung development and

kidney function remain or when specialized treatment changes lung development. A surviving child with severe kidney failure may need ongoing dialysis, tube feeding, repeated hospital care, and eventually assessment for kidney transplantation. Limb or joint differences may require physical therapy or orthopaedic care.

The course cannot be predicted from a single finding. The cause, timing of fluid loss, lung size, kidney function, prematurity, and response after birth all matter.

Follow-up after pregnancy is useful even when the baby does not survive. A review can bring together ultrasound findings, genetic results, placental examination, or post-mortem findings. This may clarify what happened and what it could mean for future pregnancies. It is also reasonable to request grief support months or years later; there is no fixed timetable for bereavement.

When to get medical help

If Potter syndrome or very low amniotic fluid has been diagnosed, keep the scheduled follow-up with the fetal medicine and pregnancy teams. Contact them between appointments if you have questions about changes in fetal movement, leaking fluid, or the birth plan.

Seek urgent pregnancy assessment if you notice:

These signs do not necessarily mean Potter syndrome is present or worsening, but they need prompt assessment.

Severe breathing difficulty in any newborn is an emergency. Call emergency services immediately if a baby is struggling to breathe, is not responding normally, or has blue, grey, or very pale skin.

Grief can also become urgent. Seek immediate crisis or emergency support if you feel unable to stay safe or are thinking about harming yourself.

- A gush or continuing trickle of fluid from the vagina
- A clear reduction in the baby's usual movements
- Vaginal bleeding
- Regular painful contractions, fever, or feeling acutely unwell

Common questions

Is Potter syndrome always fatal?

It is often fatal, especially when both kidneys are absent and the lungs are severely underdeveloped. Rare survival has occurred with specialized intervention and intensive kidney support. Outcomes may be different when the fluid loss started later or some kidney and lung function remain.

Can a baby survive without kidneys?

A baby cannot survive long-term without kidney replacement treatment. Dialysis can perform part of the kidneys' work, and transplantation may become possible later. However, treatment is only possible if the lungs and other organs can support survival.

Does low amniotic fluid always mean Potter syndrome?

No. Amniotic fluid can be low for several reasons, and mild or late-onset low fluid does not necessarily produce the Potter pattern. Potter syndrome involves severe, usually prolonged fluid loss together with effects such as underdeveloped lungs and characteristic positioning changes.

Can Potter syndrome be seen on an ultrasound?

Ultrasound can show very low fluid, absent or abnormal kidneys, bladder problems, and associated development differences. Low fluid can make scanning difficult, so repeat ultrasound or fetal MRI may sometimes be needed.

Is Potter syndrome inherited?

Not always. Bilateral renal agenesis and other kidney abnormalities may occur without a known inherited cause. Some cases are part of a genetic or chromosome condition. Testing and genetic counselling can help estimate the chance of recurrence.

Can amniotic fluid be replaced?

Fluid can be placed into the amniotic sac through a needle, but the added fluid may not remain there and does not restore kidney function. Repeated amnioinfusions are being used or studied only in highly specialized settings for carefully selected pregnancies.

Sources

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