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Background

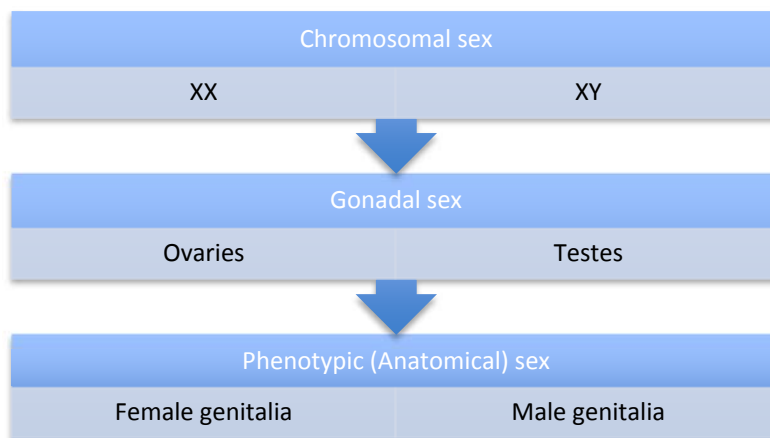
The management of a newborn with a disorder of sex development (DSD) presents a unique challenge to both health care providers and families alike. Parents report experiencing stress, confusion, grief, fear and sometimes even clinical levels of post-traumatic stress symptoms. Their initial interaction with health care providers appears to have a critical influence on their experience.

This guideline was created for the purpose of supporting health care providers caring for the newborn with a possible DSD. By following a standard local approach, we hope to ensure consistent messaging from all providers and create as smooth an experience as possible for the family. The primary management goals are to reach a diagnosis and gender assignment as soon as possible, ensure medical stability of the infant, and provide psychosocial support for the family.

**Please note: This guideline may not be applicable to all infants. Please call endocrinology if you unsure whether activation of this protocol is appropriate.**

Terminology & Classification

Disorders of sex development are defined as congenital conditions in which development of the chromosomal, gonadal or anatomical sex is atypical. They are broadly categorized as sex chromosome DSD, 46XY DSD and 46XX DSD. Many infants with these conditions present with ambiguous genitalia.

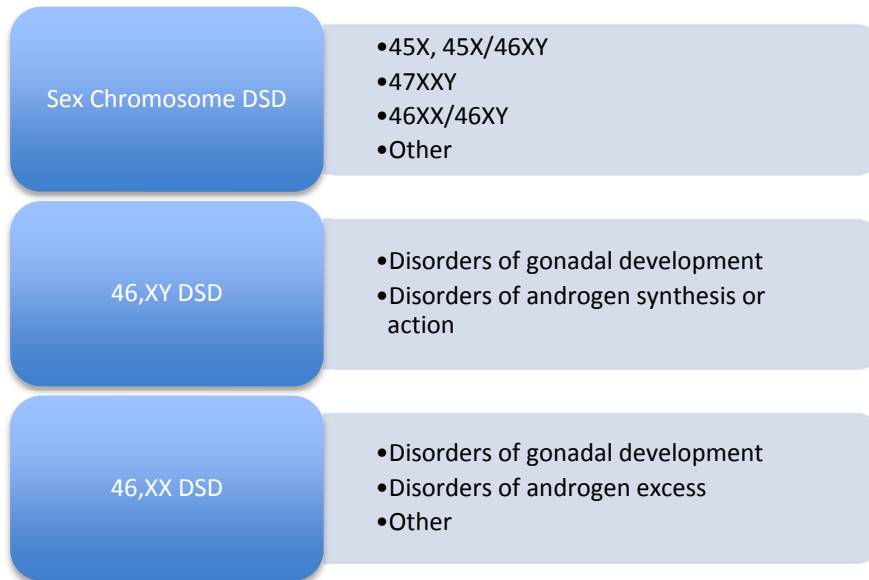


Consensus Statement on Management of Intersex Disorders.  
Pediatrics 2006; 118; e488-500.

## Approach to the Infant with Ambiguous Genitalia

Reviewed and approved by the Sections of Endocrinology, Genetics, Urology and General Pediatrics

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Initial Management Process

Ideally, when an infant is found to have ambiguous genitalia, a consistent sequence of events should take place to address the medical needs of the patient as well as the psychosocial needs of the family. Depending on the condition, this process may take several weeks to complete. It is expected that the infant will be kept in hospital for at least 1 week. In summary, the sequence of events is as follows. Details are below.



Please note: Mild cliteromegaly or isolated hypospadias may not require this protocol. These infants should be seen by a pediatrician who can determine the need for appropriate specialty consultation. Please call endocrinology if you uncertain whether the full protocol should be activated.

### When an infant is noted to have ambiguous genitalia, the responsible physician should:

- Explain their concern to the parents as soon as it has been identified
  - Not sure what to say? See [Scripts](#) for some ideas.
- Consult Neonatology right away to evaluate the infant and speak with the parents. Neonatology will facilitate consultation of additional subspecialties and, if necessary, patient transfer to ACH.
- Encourage the parents to **delay naming and gender assignment**
- Support the parent-child interaction
  - This is not a medical emergency. Parents should still be encouraged to bond with their newborn, breastfeed etc if the infant is otherwise stable.
- Use gender-neutral terms to refer to the baby.
  - Eg “your baby, the baby, your little one”
- Engage the parents while examining the baby
  - Use gender neutral terms to describe anatomy:
    - Phallus
    - Single opening/two openings
    - Labioscrotal folds
    - Gonads
  - Examining the baby with the parents can help them understand your concern and the need for further investigation. It also promotes a sense of openness rather than shame or secrecy.
  - Be sensitive about repeat examinations by multiple trainees and providers.
- Discuss the management process in simple terms, ensuring the parents are as informed as possible about what to expect. The following information should be conveyed to the parents:
  - The baby will need to remain in hospital for clinical observation, investigations and to be seen by the necessary subspecialties. **This may take a week or more.**
  - When possible, transfer to ACH may be considered to facilitate management.
  - Subspecialties involved will include neonatology, endocrine, genetics and urology. Each of these services will need to come meet the family and examine the baby.
  - Investigations will be recommended and facilitated by the consulting services. This will typically involve blood work and a pelvic ultrasound. The responsible physician should refrain from ordering investigations until advised by endocrinology and genetics.
  - Once results are available, endocrine, genetics and urology will hold a team conference to review and interpret the results. As a group they will come to an agreement on diagnosis and the recommended gender assignment, if sufficient information is available to do so (it is important to note that in certain circumstances, more extensive testing is necessary before a recommendation can be made).
  - The team will then hold a meeting with the family to review results and recommendations. Only then should the gender assignment be made.
- Offer psychosocial support
  - The unit social worker is likely the most easily accessible initial resource to help parents cope with the shock and grief associated with the news of a possible DSD.
  - Once endocrinology is involved, the endocrine clinic social worker will connect with families as early as possible to provide ongoing support.
- Discuss disclosure to friends/family
  - Parents should be encouraged to delay a formal birth announcement until a gender assignment has been made. However, they should be encouraged to be open about their current situation with their close friends/family, depending on their degree of comfort. Open disclosure with trusted individuals can help reduce parental feelings of isolation, shame and secrecy surrounding the diagnosis. The resources below can help families talk to others about their child’s diagnosis.
- Provide [reliable resources](#)

### REMEMBER

**DON'T assign a gender.** There are many factors that impact gender assignment, and these will be taken into consideration by endocrinology, genetics and urology when they meet to review results. They will then meet with the family to discuss the recommended gender assignment.

**DO identify your concern immediately.** The first provider to identify ambiguous genitalia should inform the parents of their concern right away and involve the most responsible physician for further discussion.

**DON'T disclose results as they become available.** Results will be reviewed in the family meeting with neonatology, endocrinology, genetics and urology.

### Resources for parents

- [www.aboutkidshealth.ca/En/HowTheBodyWorks/SexDevelopmentAnOverview](http://www.aboutkidshealth.ca/En/HowTheBodyWorks/SexDevelopmentAnOverview)
  - Basic information on the process of sex differentiation, with interactive visuals. Specific information on several conditions.
- [www.dsdfamilies.org/dsdfamilies/brochures.php](http://www.dsdfamilies.org/dsdfamilies/brochures.php)
  - Brochure: **“When your baby is born with genitals that look different”**. Provides basic information regarding DSD, next steps and how to communicate with friends/family.
- [www.dsdguidelines.org](http://www.dsdguidelines.org) - **“Handbook for Parents”**
  - Provides much more extensive information than may be applicable to parents at the beginning, but may be helpful early on for parents who desire more information. The first chapter is particularly helpful in the early stages of diagnosis.

### Resources for health care providers

- The above resources for parents can all be helpful for providers as well!
- [www.dsdfamilies.org](http://www.dsdfamilies.org)
- [www.dsdguidelines.org](http://www.dsdguidelines.org) - Clinical Guidelines for the Management of Disorders of Sex Development in Childhood
- Consensus Statement on Management of Intersex Disorders. Pediatrics 2006; 118; e488-500.

Scripts
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**How to notify the parents of your concern regarding a possible DSD:**

When I examined your baby, I noticed some features that are important for you to know about. What I noticed is... *[engage parents in the physical exam, describe physical findings using gender-neutral terms: phallus, opening(s), labioscrotal folds, gonads. Avoid using the term “ambiguous genitalia” with the parents].* When a baby is developing, many steps are involved to create the different structures on the inside and on the outside that make a baby look male or female. Most of the time these all come together in such a way that the sex of the baby is not in question. Sometimes a change or a variation in one of those steps causes a change in how the genitals develop, which can make it hard to tell the sex of the baby right away. It looks like this kind of change may have happened for your baby, causing *[describe physical findings]*. We will need to do some tests that will give us more information and help us understand why this happened for your baby. I wish I could tell you right now, but until we know more we really can't determine the sex of the baby. In the meantime, I would encourage you not to name your baby or to send out a birth announcement, but many parents do find it helpful to tell their close friends or family about what is happening so they can support you through this process. We can help you figure out how to share this news with them. I am going to ask the neonatologist to come and meet with you right away, so that they can speak more with you about why this may have happened and what will happen next.

**What you can say when the parents ask what to tell their friends and family:**

*(Adapted from Clinical Guidelines for the Management of Disorders of Sex Development in Childhood, 2006)*

This is important. We strongly recommend being open and honest with your close friends/family about your child's situation. Even if you don't intend to, lying or withholding information will create a sense of shame and secrecy. Though it can be awkward to talk with family and friends about a child's sex development, being honest signals that you are not ashamed—because you have nothing to be ashamed of—and it also allows those close to you to provide you with the love and support you may need. Isolating yourself at this time will probably make you feel unnecessarily stressed and lonely. Talking about it will help you feel connected with others.

In the beginning of this process, you may feel overwhelmingly emotional when you talk about your child's situation. The team will give you many opportunities to talk about your reactions and to come up with a way to share this information with family and friends. It is our experience that parents are proud of their children and do not intend to act as if they are ashamed or embarrassed by their conditions. But when they find themselves not able to openly or honestly talk about their child, over time it can magnify feelings of shame for the child. More importantly, children diagnosed with DSDs also develop feelings of shame if the topic is avoided, simplified, or continually redefined. We understand that developing these skills and establishing your comfort level will take time and support. Providing help for you in a way that is tailored to your needs and the needs of your child and family is what the team is all about.

So here is what you can tell people: “Our baby was born with a kind of variation that happens more often than you hear about. Our doctors are doing a series of tests to figure out whether our baby is probably going to feel more like a boy or a girl. We expect to have more information from them soon, and when we do we'll send out a birth announcement with the gender and the name we've chosen. Of course, as is true with any child, the various tests the doctors are doing are not going to tell us for sure who our baby will turn out to be. We're going to go on that journey together. We appreciate your love and support and we're looking forward to introducing you to our little



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one in person soon.”

It also helps to let your friends and family know whether your baby is healthy or whether there are some health concerns. Finally, take some pictures of your baby’s face and share those pictures with others!

We think you’ll encounter what other parents we’ve worked with have experienced, that family and friends usually have many questions and lots of advice. Our team of specialists will be spending lots of time with you to and will be available to answer your questions. Also, you might find it helpful to talk with our unit social worker about the emotions you may be experiencing. The endocrine clinic social worker will also be available to help you through this process once you have met the team. We know that this isn’t an easy road to walk down at first, but you’re not the first to walk down this road, and you won’t walk it alone.