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Consortium



Center for Reproductive Health After Disease

Fertility Preservation Fact Sheet

Alpha-Mannosidosis

5 Questions to Ask Your Child's Health Care Provider

1. How is Alpha-Mannosidosis affecting my child's health right now?
2. How quickly does my child need to start treatment?
3. Will treatment harm my child's future fertility?
4. What fertility options are out there?
5. Can my child have children in the future?

What is Alpha-Mannosidosis?

Alpha-Mannosidosis is a rare lysosomal storage genetic disorder. People with Alpha-Mannosidosis have gene mutations, or misspellings in the body's instructions, which stop the body from breaking down an important sugar molecule called mannose.

Alpha-Mannosidosis causes immune deficiency, which makes it difficult for the body to fight infections. Those with the most severe form of Alpha-Mannosidosis typically do not survive childhood due to repeated severe infections. Additional features of Alpha-Mannosidosis include changes in the face and bones, hearing loss, and mental retardation. Some individuals with Alpha-Mannosidosis may have problems with coordination, muscle movement, and muscle weakness.

How Does Alpha-Mannosidosis Treatment Harm Fertility?

The standard treatment for Alpha-Mannosidosis is hematopoietic stem cell transplant (HSCT). However, Alpha-Mannosidosis is rare, so there is limited data regarding how safe and effective HSCT is for people with Alpha-Mannosidosis. Before a child can have HSCT, they may have to have total body irradiation (TBI) and conditioning medication to prepare their body for the transplant. Both TBI and the necessary conditioning medications are considered *gonadotoxic*, meaning they harm the reproductive organs. A well-established risk of HSCT conditioned with TB and high doses of gonadotoxic drugs is infertility.

The Center for Reproductive Health After Disease: Alpha-Mannosidosis

Female Fertility Preservation Options

HSCT has a high risk of ovarian failure in females, which leads to premature menopause, subfertility, and infertility. Fertility preservation options should be discussed with parents of children needing HSCT for Alpha-Mannosidosis.

Girls with Alpha-Mannosidosis need HSCT as early as possible and usually have not gone through puberty at this time. Young girls' ovaries can't be stimulated, so the only option available to preserve fertility is *ovarian tissue cryopreservation*. Ovarian tissue cryopreservation involves surgically removing part or all of an ovary and freezing it until the female is ready to start a family. At that time the tissue is thawed and either transplanted back to the female to begin menstrual cycles and achieve a natural pregnancy, or the follicles are grown into eggs which are then fertilized with sperm and the embryo (fertilized egg) is implanted into the female.

Male Fertility Preservation Options

HSCT has a high risk of *azoospermia*- a medical condition where there are no sperm present in semen, therefore fertility preservation options should be discussed with parents of children needing HSCT for Alpha-Mannosidosis.

Boys with Alpha-Mannosidosis need HSCT as early as possible and have usually not gone through puberty at this time. Because pre-pubertal boys cannot produce semen, the only option available to preserve fertility is *testicular tissue cryopreservation*. Testicular tissue cryopreservation involves a biopsy to remove testicular tissue, which

contains cells that create sperm and sperm itself. The tissue is frozen until the male wishes to start a family. At that time *intracytoplasmic sperm injection* (ICSI) can be used to achieve fertilization. ICSI is when a single sperm is microinjected directly into an egg.

Alternative Options

Those with Alpha-Mannosidosis who survive into adulthood may choose to use donor sperm or eggs, or consider adoption when family planning.

Questions?

Call the 24-hour FERTLINE to ask your fertility preservation questions, get connected with a fertility preservation program near you, and access resources, tools, and support!



Online Resources

<http://oncofertility.northwestern.edu>

<http://www.myoncofertility.org>

<http://www.savemyfertility.org>



Information found here or elsewhere on the oncofertility.northwestern.edu website should not be considered medical advice, diagnosis, or treatment. Any information on this document or website should not be used in lieu of consultation with your healthcare provider or physician. Before starting any course of treatment, always consult a qualified health care provider. Do not delay seeking or disregard medical advice because of anything you have read or seen here. For information regarding fertility options contact the FERT line at 866-708-FERT (3378).