

SEX AND THE MALE WITH PWS
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In the past, our primary concern was saving our children's lives. Although this is still the number one priority, we now are also at a point where we are able to focus on the quality of their lives. One issue that has had little attention in the past is addressing the sexual functioning of our teens and young adults with PWS, and their sexual needs. One way we have addressed that at PWSA (USA) is to sponsor an extensive study in Israel by Drs. Varda Gross-Tsur and Harry Hirsh, and cooperating on another study By Drs. Susan Myers and Barb Whitman on a similar topic. We will report the results of the Israel research project which is close to completion in the near future.

We have also addressed this topic in the 21st chapter, *Advocacy Issues: Sexuality*, in our 3rd edition of the PWSA (USA) sponsored **Management of PWS** book. I was involved with writing this chapter along with David Wyatt, and Barbara J. Goff. I also wrote an article From the Kitchen to the Bedroom – Sexuality and Prader-Willi Syndrome that was published in *The Gathered View*. So, I have waded into this delicate topic before, but this time I would like to wade a little deeper, and address questions that I have received on the sexual functioning of males with PWS. I feel the people who have asked the questions – and those who are too embarrassed to ask – deserve some answers.

Questions I have received at PWSA (USA) from male adults with PWS via email:

- I have one question to ask you. Can people that has Prader Willie Syndrome can one day can they be a Parent yes or no?
- hi merry christmas can you send me info about gonadotropin please i really want to have kids with my girlfriend when i get married to her please i really do.
- Will this diseases make me sterile?
- i was wondering what is the special kind of medicine for producing sperm is it called genetrobin something like that. well i wanted to know so i can tell doctor i wanted to get married and have my own family im one of them can produce sperm i had a normal erection and i just wanted to know what was it called so i can have them stronger. Thanks. write back soon
- [Do I need sex hormone replacement therapy? What do they do to me?](#)

Questions I received from a mom:

Dear Janalee

My son with PWS got involved with a very cute girl in his village. She invited him to her bed, and alas, "it" didn't work at all. He is so miserable. I know he has erections from time to time but he himself says that it is all "dry". This time he didn't have erection what so ever. His penis is small but not tiny. Do you know if our boys can function sexually at all? I mean are they able for intercourse? I didn't believe we shall get to those corners so early, and we were caught absolutely unready.

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The following are responses from endocrinologists on our medical advisory boards:

1) The treatment in general of micropenis (small penis) for anyone is better when treatment is given during infancy or early childhood.

Early in life males have large number of androgen receptors but they gradually decrease with age, therefore late treatment with androgen can give erection but not enlargement of their penis. In theory, individual with PWS could be treated with a combination of LH and FSH as it is done in another condition with hypogonadotropic hypogonadism known as Kallmann syndrome in whom, the main characteristic beside the hypogonadism is anosmia (lack of sense of smell). Androgen replacement improves both libido and erectile function. Restoring fertility is theoretically possible in patients who generally respond to treatment with pulsatile GnRH or gonadotropins.

This is another reason why I recommend medical management with gonadotropins early in life to male infants with PWS with undescended testicles before surgery, because even in the case of failure to bring the testicles down to the scrotum, the testis still produces testosterone under such stimulation which could help for further enlargement of the penis

Androgen therapy in males should be started with low dose followed by gradual increase as tolerated to avoid behavioral problems seen at the normal recommended dose for hypogonadic males.

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2) Testosterone will not significantly affect penile length in a sexually-mature male, e.g. a male who has been previously exposed to full testosterone levels either naturally or via medication for an extended period of time. However, a male with hypogonadism (abnormally low natural testosterone production) who has not received full medical replacement therapy will have a response to testosterone therapy.

On a purely theoretical level, males with PWS should be able to correct their infertility if they receive gonadotropin replacement or if we knew of a way to restore endogenous gonadotropin secretion. The testicles are generally normal (except in cases of testicular damage due to maldescent and/or surgical damage) However, in practice, it is rather difficult to correct gonadotropin deficiency in males. This is true in other situations of male hypogonadotropic hypogonadism, not only PWS. The reasons for this are complex, but basically have to do with the need for pulsatile gonadotropin secretion, which is difficult to replicate using medications. Some andrology clinics have had limited success using gonadotropin pump therapy in cases of hypogonadotropic hypogonadism, but I am not aware of any attempts in males with PWS. We now also recognize the importance of other hormones, such as inhibin and activin, in the control of male fertility, but I am not aware of any studies specifically looking at these hormones in PWS.

Phillip Lee, M.D.