Anre's Story

Congenital Adrenal Hyperplasia

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There are many ways to be Intersex; one of the most common is to have Congenital Adrenal Hyperplasia (C.A.H.). There are two main types of C.A.H., 21 Hydroxylase and 11 Beta Hydroxylase, the first accounting for about ninety percent of all C.A.H.. 11 Beta accounts for five to six percent. There are several other kinds of C.A.H. but they are rare.

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Both 21 Hydro and 11 Beta can cause higher than normal production of androgens in the adrenal gland and consequent virilisation (masculinisation). C.A.H. is generally caused by a failure to produce one or more of the hormones usually produced in the adrenal gland. When those hormones are not produced, their precursors form testosterone.

Only 21 Hydro C.A.H. is associated with life threatening illness. In 21 Hydro, two critical-to-life hormones are, to a greater or lesser extent, absent, so that the body does not process salt ('salt wasting'). The inability to retain salt leads to "adrenal crisis" that if left untreated can be fatal. Prior to the discovery of mineral corticol replacement therapy, children born with severe 'salt wasting' usually died within weeks of birth. With other forms of C.A.H. the most significant feature is ambiguous genitalia or other forms of virilisation such as hair distribution, skeletal structure and muscle mass.

Anre was born in the early 1950s in a small rural town several hours drive from the nearest capital city's specialist children's hospital. Anre was born with 11 Beta C.A.H. and was so significantly virilised that doctors attending the birth were unsure of Anre's sex.

Apart from ambiguous genitals there was no other apparent difference or illness at the time of Anre's birth. For reasons that are now impossible to determine, even through legal process, Anre was assigned female even though neither genetic testing nor internal inspection were possible at the small rural hospital in which Anre was born.

Despite the lack of illness, doctors were so concerned at Anre's appearance that they advised Anre's mother to attend the distant children's hospital as soon as possible. Medical experts there were unsure of the cause of Anre's differences so they contacted the then world-leading specialist hospital on Intersex differences, Johns Hopkins in Baltimore, Maryland, U.S.A.. The advice from Johns Hopkins was extraordinary.

Johns Hopkins was at that time, in the process of first describing 11 Beta C.A.H.. They knew that, left alone, Anre's adrenal gland would continue to produce testosterone at male levels and Anre would continue to virilise. They also knew that Anre would experience precocious puberty, short stature and skin pigmentation. Johns Hopkins advised Anre's specialists to remove one adrenal gland, take a part of that gland and retransplant it in Anre's upper thigh. If the transplanted section was able to produce sufficient mineral corticols then the remaining adrenal gland should also be removed.

Anre was three by the time the surgery was performed. While the adrenal surgery was being performed doctors decided that it was an appropriate time to conduct genital reconstructive surgery. The reasons for the decision to assign female and to confirm that with surgery cannot be determined.

Anre in later life commenced legal proceedings to recover as many of the pertinent medical documents as possible. Although the nature of the surgery and the specialists' advices were discoverable, there is no evidence of genetic testing or internal examination prior to assignment. Anre supposes that sufficient evidence of a vagina, the position of the urethra and the relative ease of surgical assignment as female were the reasons.

At the time medical assignments of Intersex were more often male than female. There were two fundamental reasons for this: The first was the relative infancy of plastic surgery and uncertainty about outcomes. As surgical technique improved through the 1950s and '60s practitioners were more and more likely to recommend definite assignments that could be assured with surgery. At the same time theories about the plasticity of gender in infants were being proposed so that, it was thought, irrespective of innate anatomy, a child could be raised to believe they were boy or girl by social conditioning. The second was the then prevailing Victorian-era notion of male privilege. It was thought that any child would be best advantaged by a male assignment so that wherever possible children whose sex seemed ambiguous should be treated and raised as male. This notion of male privilege is still prevalent in most countries. Complexities around surgical assignments, where fully functioning genitals are the goal, makes female assignments technically easier so that, these days the desire to bestow male privilege takes second place to ease of surgical assignment.

For Anre the surgery performed was not a success. The genital reconstruction surgery was little more than a clitorectomy. Surgical technique did not then extend to vaginal reconstruction. Anre's adrenal transplant failed so only one functional gland remained. This

was sufficient to provide mineral corticols and continue Anre's virilisation unabated.

By age nine Anre was entering puberty, as predicted by Johns Hopkins. Anre was also exhibiting physical attributes inconsistent with the assignment of female. Further consultations with Anre's Australian specialists, and those who were closely following the case from Johns Hopkins, resulted in a recommendation for further adrenal gland reduction. This time it was thought prudent to leave the gland where it was and to surgically reduce its size (resection). No further attempts were made at genital reconstruction although painful dilation was recommended as a way of increasing the size of Anre's tiny vagina.

The need to be close to the children's hospital for Anre's many medical check-ups caused Anre's mother to decide to move in to town. This move, coupled with news that Anre's differences were most likely passed down from one of the parents, caused an irreparable rift between Anre's mother and father who then separated.

Both parents need to be recessive carriers for the child to be affected by this form of C.A.H.. This was unknown to Arne's specialist.

Although the so-called sex chromosomes X and Y were discovered by Stevens and Wilson in 1903, testing was difficult and seldom conducted. D.N.A. itself was not described until 1953 by Crick and Watson so that a precise understanding of genetic causes for human differences was still many years in the future. So far as Anre's father was concerned the mother was the problem and Anre reports "the doctors didn't disagree".

Anre recovered from the adrenal section surgery but the resection was so extensive that satisfactory production of mineral corticoids (a class of steroid hormones characterised by their influence on salt and water balances) could not be achieved. Anre was discharged with monthly, then annual follow-ups. Doctors became concerned at low levels of mineral corticoids and prescribed a regime of replacement therapy, then only just coming into use. The long-term consequence of this medication was unknown so Anre was part of a study group measuring the side-effects of the new drugs.

By the age of fifteen, Anre was being prescribed dexamethasone. The side-effects of this drug are quite profound. Weight gain and mood swings made Anre a reluctant user and every opportunity to avoid the drug was taken. Anre retained sufficient adrenal function to, for the most part, get by without chemical assistance. Anre's inconsistent use of the drugs prescribed caused considerable angst amongst medical specialists both at Johns Hopkins and in Australia. Anre could not be convinced to consistently take drugs and so was dropped from the study group. What followed was a lifetime of struggle with increasing suspicion and hostility between Anre and the specialists. Over time, endocrinologists retired or left the hospitals Anre attended so that the whole process began anew with each replacement.

Anre attended school, achieving normal education levels and enjoyed a happy childhood with mother and siblings.

At about the age of twenty-three, Anre decided to forgo dexamethasone permanently and lived drug free for twenty years. In that time Anre lived a normal life working in chemical engineering for a major Australian company.

I met Anre as an adult of roughly my own age and wondered how Anre had managed adolescence and the twenties, years when most people find a partner and settle down to family life. Anre told me, with much regret and blame to those who had attempted to normalise, that a sex life was impossible. The surgery performed essentially removed Anre's penis/clitoris and left non-functioning genitals. Anre retold conversations had with mother about the kind of life that lay ahead. That life would not include a partner. Both Anre and Anre's mother remained single and lived together until Anre's mother died recently.

From time to time Anre became ill, often for reasons unconnected with C.A.H.. Doctors nonetheless took every opportunity to convince Anre to return to dexamethasone treatment even when it was obvious that there were no symptoms of mineral corticoid deficiency. On more than one occasion Anre was threatened with psychiatric sectioning because of this refusal. Anre's mother was a vigorous and strong supporter of Anre's stand and opposed any attempts to characterise Anre as 'mad'.

To this day Anre has a strong and reasonable fear that doctors may force medication irrespective of consent or need.

By the age of forty-five, Anre's adrenal gland could no longer produce adequate hormones. Two factors were the cause: the original reduction surgery - especially the scarring that resulted and the side-effects of dexamethasone which, over time, suppresses adrenal function, long-term continuous use of dexamethasone then commenced which resulted in complete failure of Anre's adrenal gland and medically induced Addison's disease. (See iatrogenic illness)

The side-effects Anre can now look forward to are increased bone fragility, Diabetes mellitus, psychological lability, glaucoma, muscular atrophy, skin disease and hypertension. Interestingly hypertension is the single most dangerous consequence of 11 Beta C.A.H.. All of the other consequences: genital differences, short stature, skin pigmentation and infertility cause no illness. The medication that Anre was prescribed recreated the very thing any intervention should have prevented.

The sole purpose of all of Anre's surgery and medical interventions was to prevent virilisation and reverse what virilisation had taken place in what was thought to be a girl and later a woman. The interventions were not for the good of the client. They were designed to erase differences of sex within a society that found those differences repulsive. That they were not concerned with Anre is demonstrated by the specialists' absolute refusal to engage in a dialogue to determine how Anre felt about sex, gender or the surgeries and their deafness to Anre, then and now, when concerns were voiced.

Anre is now in the late fifties of life. Just what awaits so far as aged care and the consequences of the many years of dexamethasone replacement therapy is a cause of great anxiety. Anre's mother, the great defender of Anre's rights, is no longer there to argue and protect. Anre's extended family is now distant, old and has little understanding of Anre's special situation. The fear of medical practitioners is high, with several in recent times hinting at mental instability because Anre insists on seeing and interpreting test results and on having copies for private records. On occasions Anre has insisted doctors' notes be destroyed when they have made personal,

insulting observations and once has had the Australian Medical Association intervene on matters of privacy and consent.

Anre now lives quietly with two dogs and an amazingly encyclopaedic understanding of endocrinology. Anre's life shows what can happen when medical practitioners intervene, not only without consent but against every ethical precaution one would imagine being exercised when the whole of an individual's life could be ruined by experiments gone wrong.

It is easy to think that the kinds of things that happened to Anre are now in the past and were a consequence of imprecise medicine and the budding understanding of genetics and endocrinological differences such as C.A.H.. Sadly this is not the case.

Medical practitioners have now abandoned Intersex as a term that describes physical differences of sex anatomy. In its stead they use a new term 'Disorders of Sex Development' (D.S.D.). This terminology applies to people with different sex anatomies even when there is no illness associated with that difference. Differences such as Anre's where one might live a full and contented life without any medical intervention whatsoever, as happened until plastic surgery was raised to its current levels of proficiency, are now routinely subjected to genetic scrutiny and surgical intervention so that the individual is placed unquestioningly on one side or the other of the sex binary divide. Unquestioningly, that is, until the child becomes an adult and rejects the decisions that have been made without their consent.

The current protocols for the management of C.A.H. recommend early surgical intervention for children like Anre. Experts claim the surgery is much improved. They are reluctant to admit all surgeries leave scar tissue and scar tissue is insensate.

They are reluctant to ask their patients what kind of sex life they enjoy and ignore studies that say adults that had surgeries as children do not enjoy sex. They are reluctant to admit that no single surgery will resolve Intersex differences and many surgeries will be necessary.

They are embarrassed to admit that vaginal reconstruction requires constant dilation to prevent scar tissue closing up the wound, dilation of three- and four-year-old children on a daily basis.

They are reluctant to admit they have done no long-term follow-ups on their clients, indeed they make no attempt to keep track of them and have no idea what kind of life they have left for their patients to sort out.

They still conduct unethical experiments on Intersex children and proceed with surgeries without full and informed consent primarily from the child, but even the parents are not put in touch with other Intersex so they can know what lies ahead for their child if they should proceed with surgery.

Doctors test children as young as five with vibrators to see if their surgeries are successful and advise pregnant women who may be carrying a C.A.H. child to take dexamethasone to prevent ambiguous genitalia and thus, potential same-sex attraction. They give this advice despite the potentially horrific side-effects to both mother and child and before it is known for certain the foetus is C.A.H..

Medicine still performs these outrages on Intersex without reference to courts of law or ethical committees. Compare this with the experience of a transgender child who is in need of hormone blockers. It is routine for that child to be required to make application to the Family Court for permission to undertake such a drastic and life altering decision. Adrenal resections for C.A.H. children are still common and some parents go so far as allowing full adrenalectomies. Such surgeries do nothing to alleviate the symptoms of C.A.H. indeed adrenalectomies also remove adrenalin production. The target of such surgeries is testosterone and the prevention of virilised children, especially those supposed to be girls.

C.A.H. specialists and parents constantly refer to their XX children as C.A.H. girls, they refuse to acknowledge Intersex, they exclude C.A.H. adults who are Intersex and they moderate their Internet notice boards to prevent talk about Intersex. In recent discussions about dexamethasone in the *Hastings Journal* and mainstream media no mention is made of Intersex differences despite the intention of the medication being the prevention of just that.

The main C.A.H. support group in America, run by parents and doctors, supports the administration of dexamethasone to pregnant women and recommends their members to their clinics. They have helped distribute research questionnaires, on behalf of Dr. Sheri Berenbaum, on tomboy and lesbian behaviour in C.A.H. children and offered spirited defences of Dr. Dix Poppa's procedures that include the sexual stimulation of five-year-olds.

Clearly Intersex C.A.H. children cannot depend on medicine or even their parents to guarantee their rights to genital autonomy. Clearly only full human rights protection in legislation and law for those of us born with anatomical differences of sex will bring an end to these disgraceful practices.

Anre's Story was written by Gina Wilson, National President of O.I.I. Australia.

The Organisation Internationale des Intersexué (O.I.I.) is the world's largest intersex organization with members representing almost all known intersex variations. O.I.I. affiliates in twenty countries, on six continents, speaking ten languages. Visit the O.I.I. website.

Likewise, the Congenital Adrenal Hyperplasia Support Group Australia Incorporated provide support nationally for: parents of newly diagnosed children with C.A.H.; ongoing family needs and education; and adults living with C.A.H.. Visit the C.A.H.S.G.A. website.

under the S.A.A.P. program and supported by the N.S.W. Health Department through the AIDS and Infectious Diseases Branch. Polare provides a forum for discussion and debate on gender issues. Unsolicited contributions are welcome, the editor reserves the right to edit such contributions without notification. Any submission which appears in Polare may be published on our internet site. Opinions expressed in this publication do not necessarily reflect those of the Editor, The Gender Centre Inc., the Department of Family & Community Services or the N.S.W. Department of Health.

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