INTERSEX : PANDANGAN BIOETIK ISLAM DAN HUMAN DIGNITY

SAA

Ilustrasi

- Kasus Dorce yang menjalani operasi ganti kelamin
- Membedakan transgender dan transeksualitas serta interseksualitas

Defining Sex and Gender

Gender identity (Psychological sex)

Inner sense of owns maleness / femaleness.

- Sex of rearing
- Gender role

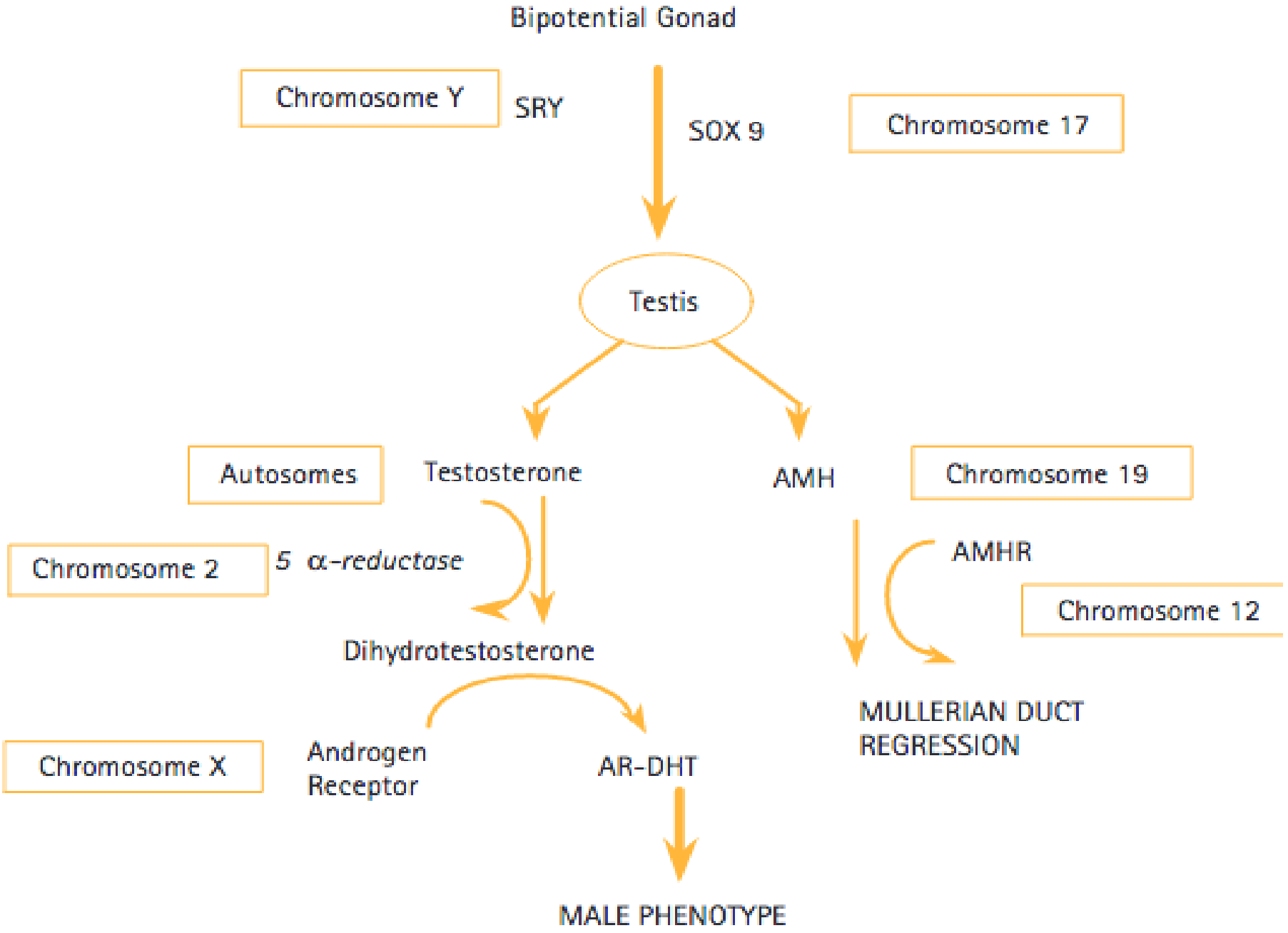
Sexual identity (Organic sex)

The biologic sexual differentiation

- Chromosomal sex
- Gonadal sex
- Internal genital sex
- External genital sex
- Hormonal sex

What is DSD/intersex?

- Disorder of sexual differentiation is the terms used for a child born without clear male or female phenotype. The term “hermaphrodite” is derived from Greek mythological God “Hermaphroditos” son of Hermes and Aphrodite, whose body after being merged with nymph Salmakis assumed a more perfect form with both male and female attributes


Bipotential Gonad
Chromosome Y
SRY
SOX 9
Chromosome 17
Testis
Autosomes
Testosterone
AMH
Chromosome 19
Chromosome 2
5 α-reductase
AMHR
Chromosome 12
Dihydrotestosterone
MULLERIAN DUCT REGRESSION
Chromosome X
Androgen Receptor
AR-DHT
MALE PHENOTYPE

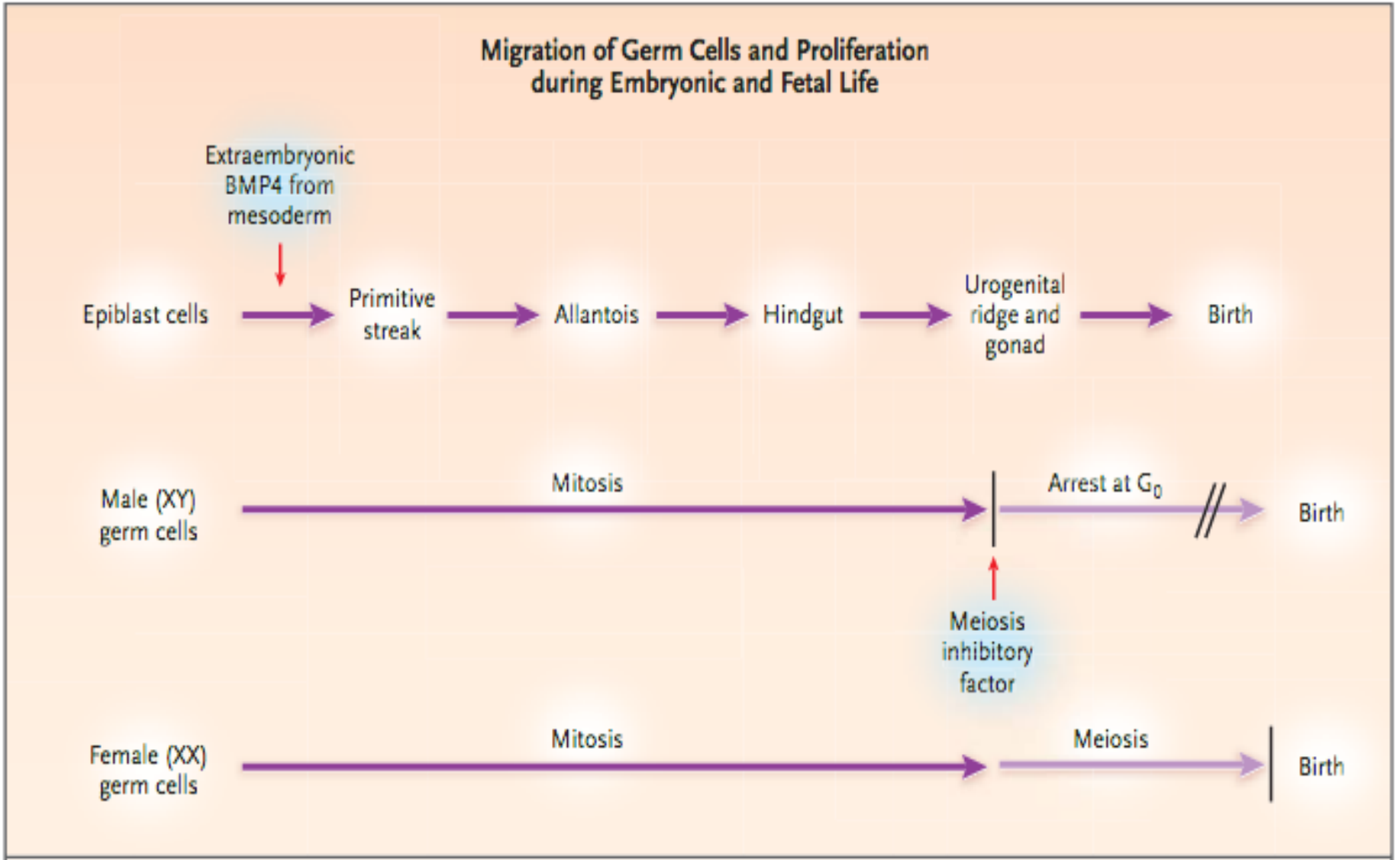


Figure 2. Migration and Proliferation of Germ Cells during Embryonic and Fetal Life.

Germ cells are first detected in the epiblast, where they are activated by bone morphogenetic protein 4 (BMP4) from the extraembryonic ectoderm. Migration occurs through the primitive streak to the base of the allantois, where alkaline phosphatase–positive cells can be detected. Subsequently, the cells migrate to the urogenital ridge, where the gonads form. The 46,XY and 46,XX germ cells undergo different patterns of mitosis and meiosis.

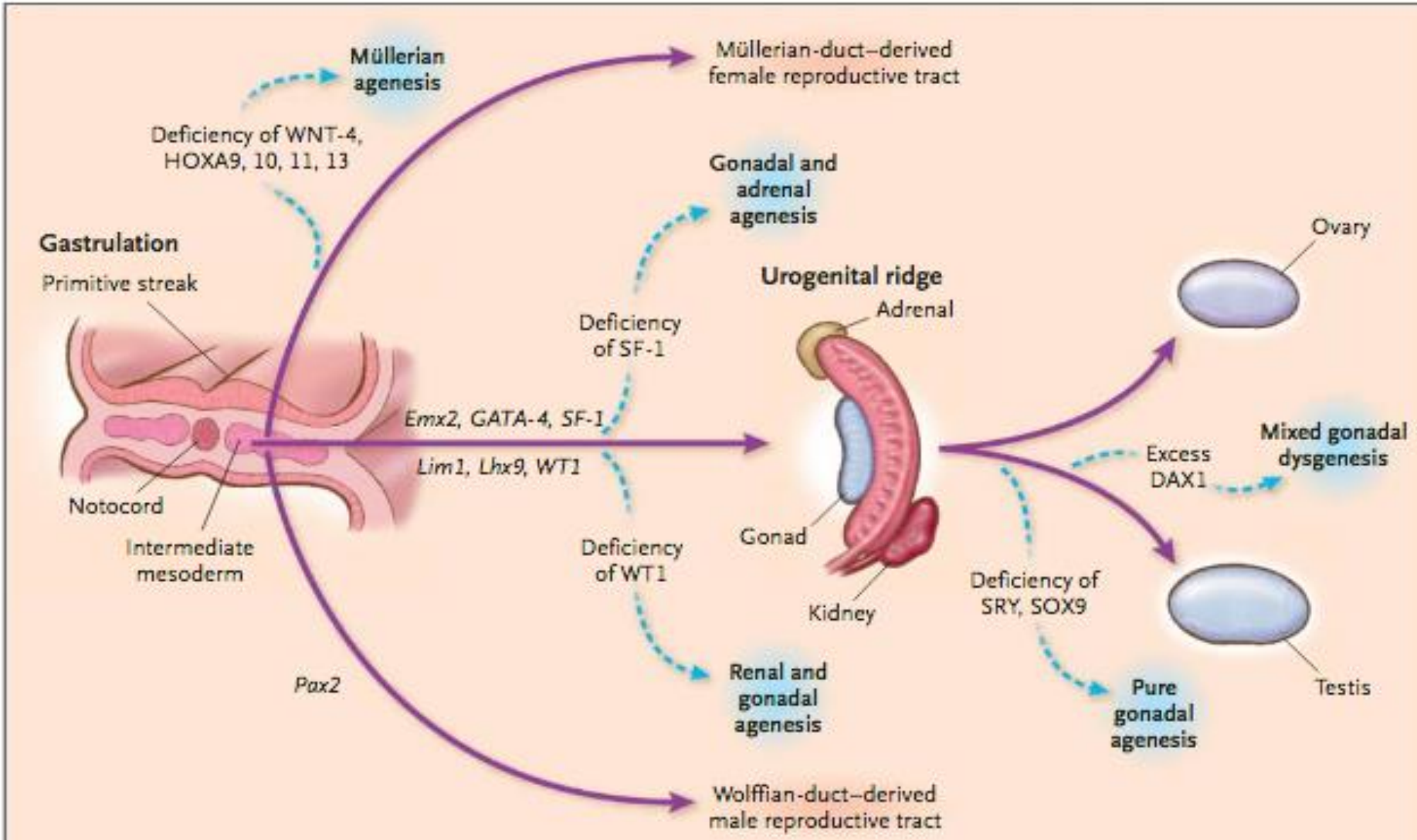


Figure 3. Syndromes of Dysgenesis during the Development of the Urogenital Ridge.

Mutations in various genes can lead to a variety of syndromes of dysgenesis involving the müllerian or wolffian ducts, gonads, kidneys, and adrenal glands as a result of a deficiency or excess of the proteins shown. *DAX1* denotes the gene for duplicated in adrenal hypoplasia congenita on the X chromosome 1; *Emx2* the empty spericles homeobox gene 2; *GATA-4* the gene encoding a protein that binds to a GATA DNA sequence; HOXA homeobox protein; *Lim1* a homeobox gene important for limb development; *Lhx9* a lim homeobox family member; *PAX2* paired box homeotic gene; *SF-1* the gene for steroidogenic factor 1; SRY sex-determining region of the Y chromosome; SOX9 SRY homeobox 9; Wnt-4 a protein that induces the development of the müllerian mesenchyme; and *WT1* Wilms' tumor-suppressor gene 1.

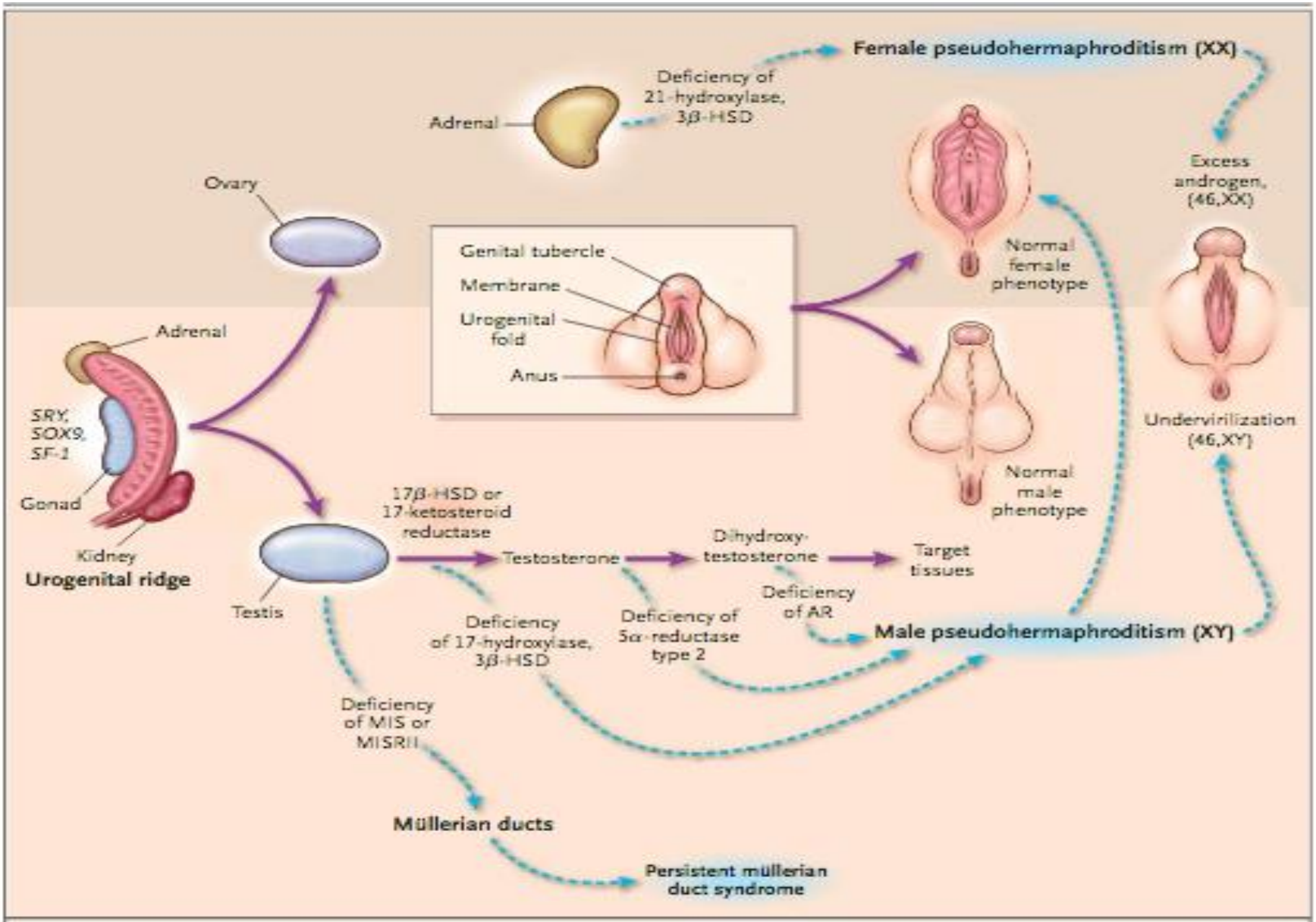
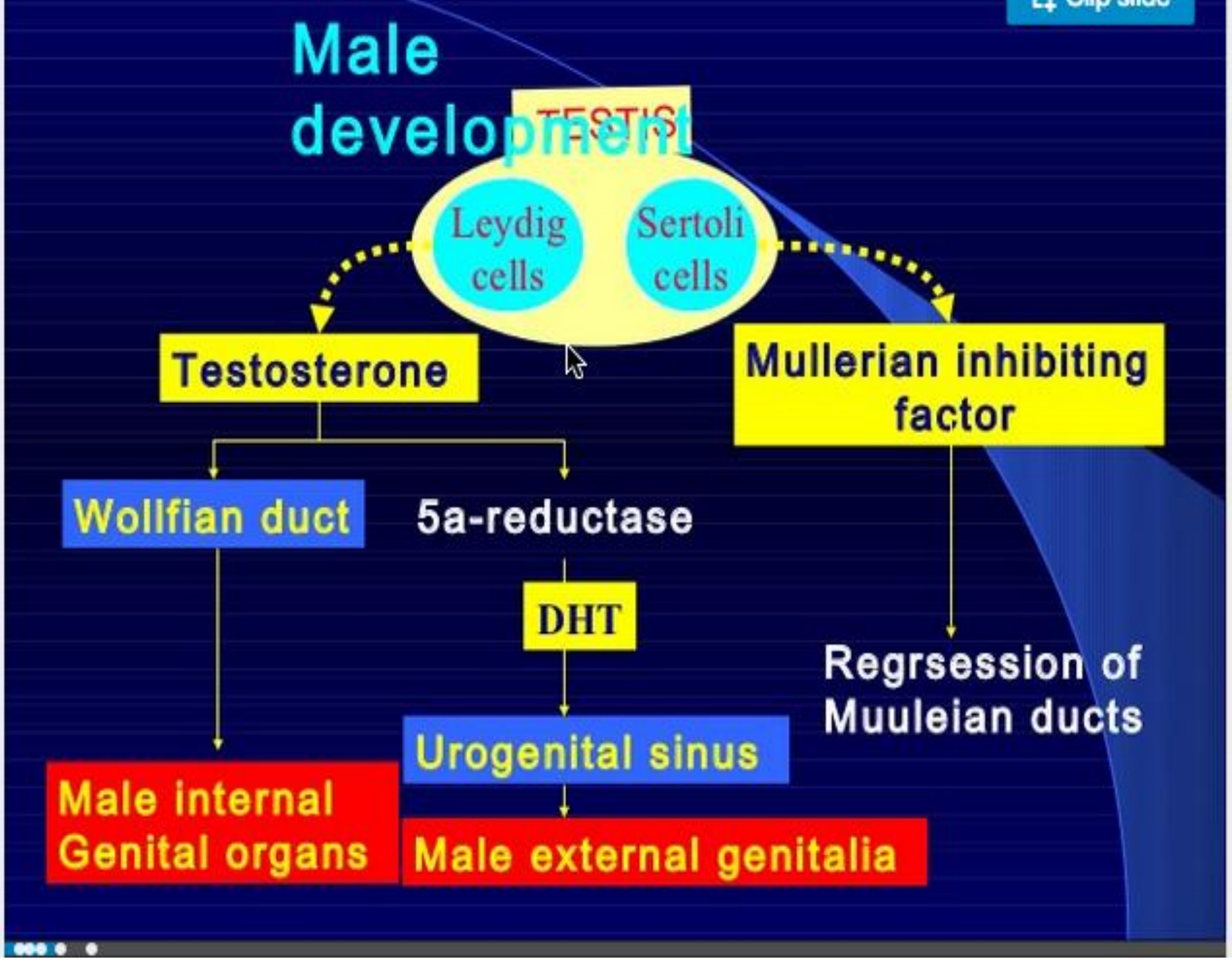


Figure 4. Functional Abnormalities of the Synthesis and Action of Hormones.

After the gonads have formed, reduced hormonal activity or signaling of specific receptors can lead to functional abnormalities of the reproductive tract, including persistent müllerian duct syndrome; male pseudohermaphroditism, causing undervirilization; and müllerian agenesis. After adrenal development, reduced enzymatic activity can result in female pseudohermaphroditism with excessive virilization. HSD denotes hydroxysteroid dehydrogenase, MIS müllerian inhibiting substance, MISRII müllerian inhibiting substance type II receptor, *SF-1* the gene for steroidogenic factor 1, *SRY* the gene for the sex-determining region of the Y chromosome, *SOX9* the gene for SRY homeobox 9,


Male development
TESTIS
Leydig cells
Sertoli cells
Testosterone
Mullerian inhibiting factor
Wollfian duct
5a-reductase
DHT
Regrsession of Muuleian ducts
Urogenital sinus
Male internal Genital organs
Male external genitalia

Female development

Development

OVARY

Urogenital sinus

Mullerian ducts

Female external genitalia
. Lower part of vagina

Absence of androgen exposure

Female internal genital Organs
. Most of upper vagina

. Cervix and uterus
. Fallopian tubes

Table 1 Proposed revised nomenclature

Previous	Proposed
Intersex	**Disorders of sex development** (DSD)
Male pseudohermaphrodite Undervirilisation of an XY male Undermasculinisation of an XY male	46,XY DSD
Female pseudohermaphrodite Overvirilisation of an XX female Masculinisation of an XX female	46,XX DSD
True hermaphrodite	Ovotesticular DSD
XX male or XX sex reversal	46,XX testicular DSD
XY sex reversal	46,XY complete gonadal dysgenesis

Table 2 An example of a DSD classification

Sex chromosome DSD	46,XY DSD	46,XX DSD
(A) 45,X (Turner syndrome and variants)	**(A) Disorders of gonadal (testicular) development** 1. Complete gonadal dysgenesis (Swyer syndrome)	**(A) Disorders of gonadal (ovarian) development** 1. Ovotesticular DSD 2. Testicular DSD (eg, SRY+, dup SOX9)
(B) 47,XXY (Klinefelter syndrome and variants)	2. Partial gonadal dysgenesis 3. Gonadal regression 4. Ovotesticular DSD	3. Gonadal dysgenesis
(C) 45,X/46,XY (mixed gonadal dysgenesis, ovotesticular DSD)	**(B) Disorders in androgen synthesis or action** 1. Androgen biosynthesis defect (eg, 17-hydroxysteroid dehydrogenase deficiency, 5α reductase deficiency, StAR mutations	**(B) Androgen excess** 1. Fetal (eg, 21-hydroxylase deficiency, 11-hydroxylase deficiency) 2. Fetoplacental (aromatase deficiency, POR)
(D) 46,XX/46,XY (chimeric, ovotesticular DSD)	2. Defect in androgen action (eg, CAIS, PAIS) 3. LH receptor defects (eg, Leydig cell hypoplasia, aplasia) 4. Disorders of AMH and AMH receptor (persistent mullerian duct syndrome)	3. Maternal (luteoma, exogenous, etc)
	(C) Other (eg, severe hypospadias, cloacal extrophy)	**(C) Other** (eg, cloacal extrophy, vaginal atresia, MURCS, other syndromes)

While consideration of karyotype is useful for classification, unnecessary reference to karyotype should be avoided; ideally, a system based on descriptive terms (for example, androgen insensitivity syndrome) should be used wherever possible.
AMH, anti-mullerian hormone; CAIS, complete androgen insensitivity syndrome; DSD, disorders of sex development; MURCS, mullerian, renal, cervicothoracic somite abnormalities; PAIS, partial androgen insensitivity syndrome; POR, cytochrome P450 oxidoreductase.

Summary of Normal Sex Differentiation

- genetic sex is determined at fertilization.
- testes develop in XY fetus, ovaries develop in XX fetus.
- XY fetus produces MIS and androgens and XX fetus does not.
- XY fetus develops Wolffian ducts and XX fetus develops Mullerian ducts.
- XY fetus masculinizes the female genitalia to make it male and the XX fetus retains female genitalia.

INTERSEX

An individual in whom there is discordance between chromosomal, gonadal, internal genital, and phenotypic sex or the sex of rearing

- **INTERSEXUALITY:**

  Discordance between any two of the organic sex criteria
- **TRANSSEXUALITY:**

  Discordance between organic sex and psychological sex components

& Kisah Hidup Dorce

1970
(usia 7 tahun)
Mulai merasakan kejanggalan dalam dirinya. Merasa keperempuan-perempuanan

1973
(usia 10 tahun)
Gabung dengan Bambang Brothers

1976
(usia 15 tahun)
Cinta pertama dengan pria asal Padang yang tinggal di Jakarta

1981
(usia 18 tahun)
Gabung dengan Fantastic Dolls, band beranggotakan para waria

1986
(usia 23 tahun)
Operasi kelamin

1986/1988
Sah jadi perempuan dengan nama Dorce Ashadi

Gamalama adalah gunung yang mendominasi tanah Ternate.

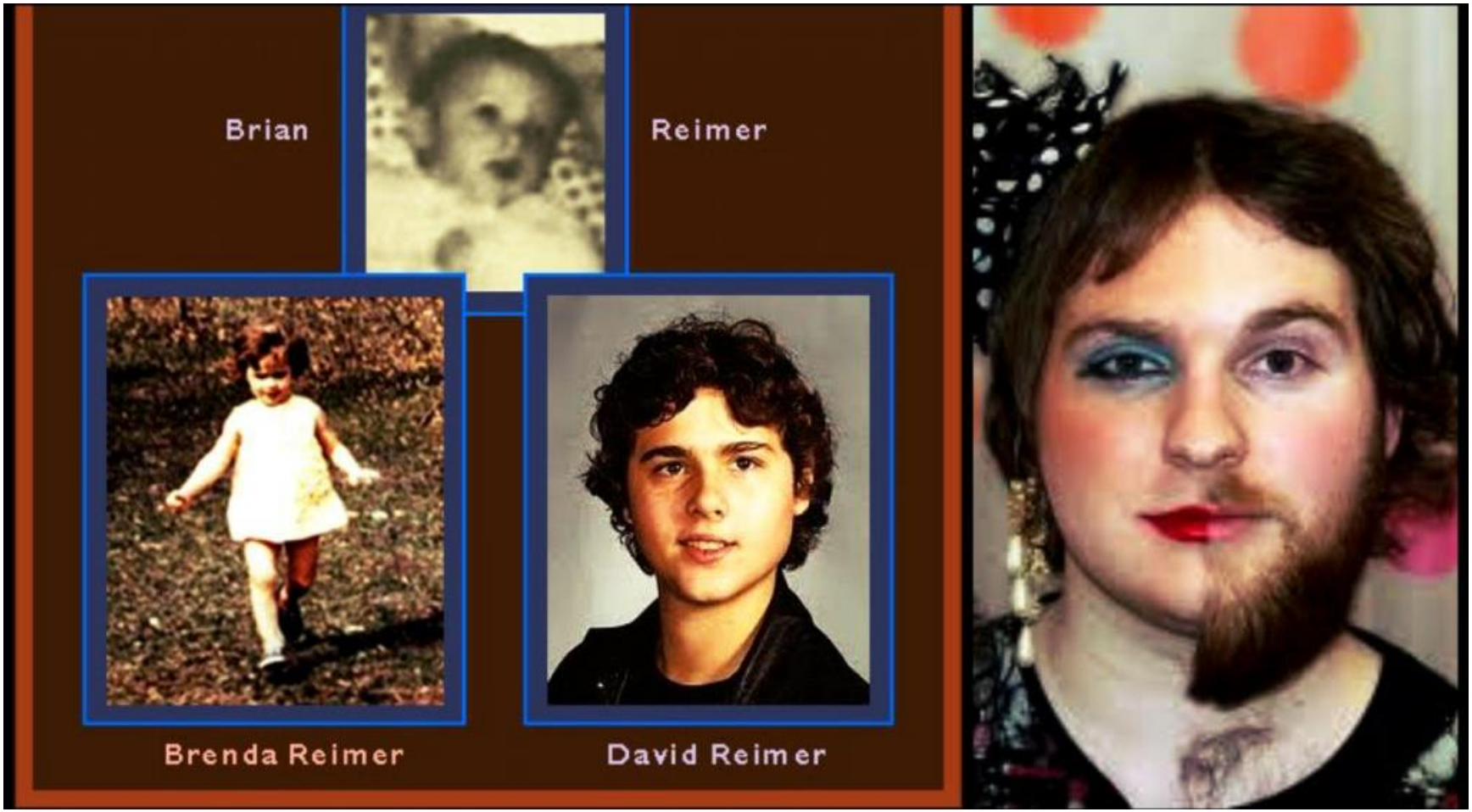
KapanLagi.com

1986
Menikah dengan Mr X
Mengadopsi seorang anak, Rizky
Pemberian nama Gamalama Oleh Sultan Ternate Mudafarsyah.

1988
Cerai

1991
Menunaikan haji lagi dan memutuskan berhijab. Membangun yayasan Dorce Halimatussa'diyah

Ilustrasi Kedua
John/Joan Case



Botched Circumcision

- **David Peter Reimer** (born **Bruce Peter Reimer**; 22 August 1965 – 4 May 2004) was a Canadian man born male **but reassignned** as a girl and raised female following medical advice and intervention after his penis was accidentally destroyed during a botched circumcision in infancy

John Money

- Money was a prominent proponent of the "theory of gender neutrality"—that gender identity developed primarily as a result of social learning from early childhood and that it could be changed with the appropriate behavioural interventions

DOGMA ILMIAH

- POSTULATE 1: INDIVIDUALS ARE PSYCHOSEXUALLY NEUTRAL AT BIRTH
- POSTULATE 2: HEALTHY PSYCHOSEXUAL DEVELOPMENT IS INTIMATELY RELATED TO THE APPEARANCE OF THE GENITALS

Ilustrasi

- www.youtube.com/watch?v=vDO24GcBcxI
- www.youtube.com/watch?v=i6HaVYg6kB4
- www.youtube.com/watch?v=WwuRSS2whTg
- www.youtube.com/watch?v=DTgHW4Hxq1w

DEFINISI MANUSIA

- HAK ASASI
- HUMAN DIGNITY
- MAHLUK DWI DIMENSI
- BERTUHAN BERSOSIAL DAN BERKEPRIBADIAN SERTA UNIQUE DENGAN ADANYA OTONOMI DENGAN KAPASITAS DAN KOMPETENSI MORAL SERTA RASIONAL
- MEMILIKI IDENTITAS JENIS KELAMIN

SIAPA YANG PALING MULIA

- LAKI LAKI ATAU PEREMPUAN YANG BERTAQWA
- TIDAK DISEBUTKAN SEPARO LAKI LAKI DAN SEPARO PEREMPUAN, atau DSD atau Intersex
- HERMARPRODITE BAGAIMANA?
- MENJADI ISU TRANSDENDER DAN HOMOSEXUALITY SELAIN TRANSSEXUALITY DAN INTERSEXUALITY
- PILIHANNYA HARUS DITENTUKAN MAU JADI LAKI LAKI ATAU PEREMPUAN
- FUNGSI REPRODUKSI LAKI LAKI DAN PEREMPUAN JELAS BERBEDA

MASALAH

- Kapan bayi dikatakan laki laki dan perempuan?
- Jika tidak jelas kelaminnya dari luar, bagaimana cara menentukan kelaminnya?
- Lantas apa pertimbangannya bayi itu akan dijadikan laki laki atau perempuan dengan tindakan medis yang sesuai?
- Apa norma agama dan hukum serta deskripsi science dan psikologis serta sosial tentang sex dan gender?
- Islam memiliki hukum waris utk perempuan dan laki laki dan imam untuk laki laki bagi sholat jumat?
- Kapan sebaiknya dilakukan intervensi medis untuk penentuan sexualitas?
- Bagaimana dengan orientasi seksualnya dan psikologisnya serta penerimaan agama dengan status perubahan kelamin?
- Apa teori etik atau bioetik yang relevan dengan tindakan medis utk perubahan kelamin atau aspek etika dengan pengakuan jenis kelamin ketiga yang apa adanya tanpa ada tindakan medis?
- Apakah perlu dirumuskan hak hak baru bagi penderita Intersex untuk menikah, warisan dan tata cara ibadah serta aturan khusus bagi mereka dengan undang undang yang sesuai?

CASE REPORT

OPEN ACCESS

True Hermaphrodite: A Case Report

Muhammad Zafar Iqbal,* Mazhar Rafee Jam, Muhammad Saleem, Mushtaq Ahmad

ABSTRACT

True hermaphrodite is one of the rarest variety of disorders of sexual differentiation (DSD) and represents only 5% cases of all. A 3-year-old child presented with left sided undescended testis and penoscrotal hypospadias. Chordee correction was performed 18 months back, elsewhere. At laparotomy Mullerian structures were present on left side. On right side testis was normally descended into the scrotum.

Key words: True hermaphrodite, Persistent mullerian duct syndrome, Disorders of sexual differentiation

APA ISU BIOETIKNYA?

- True hermaphrodite is one of the rarest variety of disorders of sexual differentiation (DSD) and represents only 5% cases of all. A 3-year-old child presented with left sided undescended testis and penoscrotal hypospadias. Chordee correction was performed 18 months back, elsewhere. At laparotomy Mullerian structures were present on left side. On right side testis was normally descended into the scrotum.

ISU UTAMA

- The birth of a child with ambiguous genitalia constitutes a social emergency
- Vulnerable
- Fragile bound

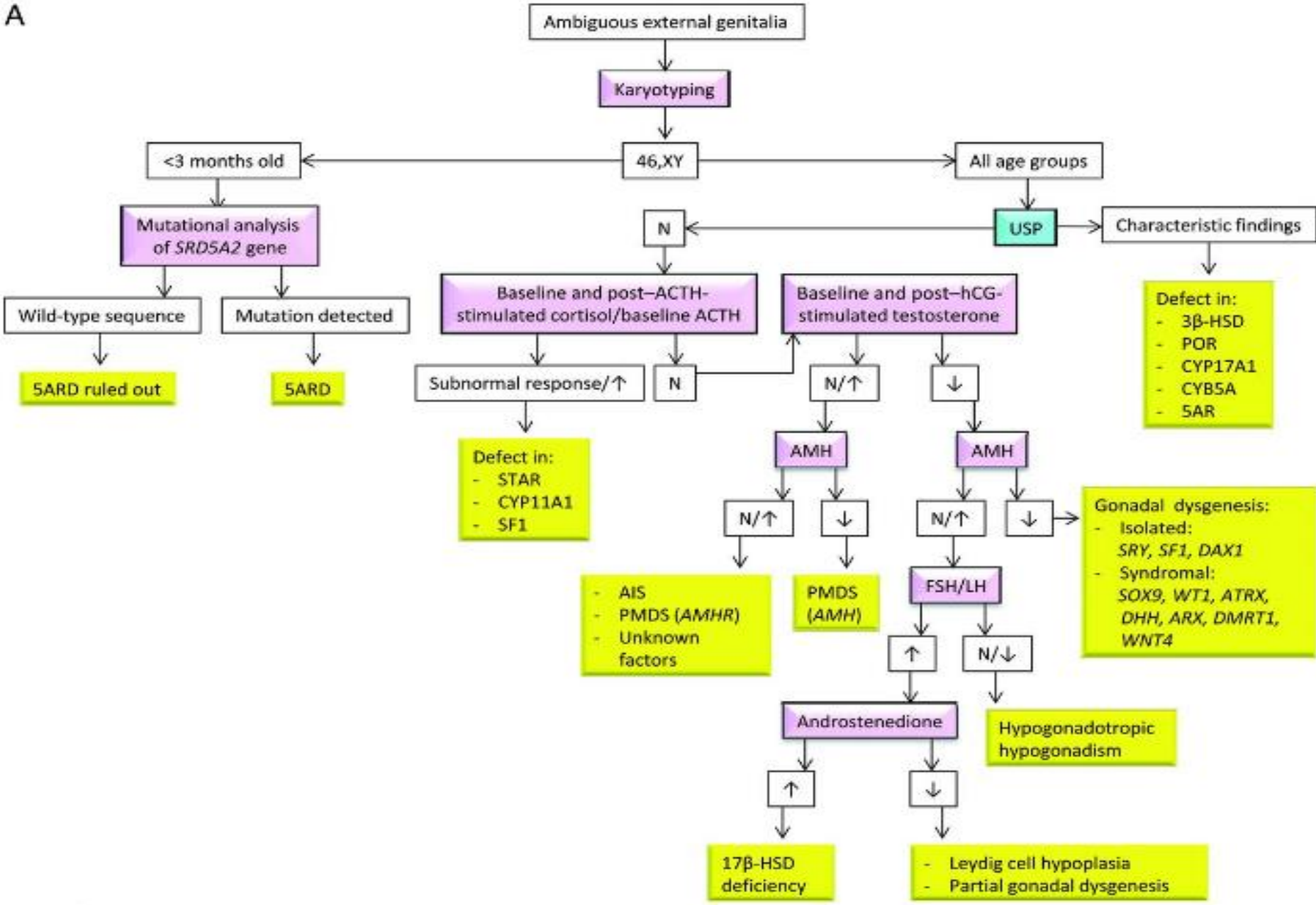
DISKUSI

- Pandangan Bioetik Islam dengan maqosidus syariah dan qowaidushariah (MS dan QS), ta'lili
- Abuse of knowledge operasi bedah plastik, bedah urology dan bedah kandungan untuk tujuan yang menyimpang dan melawan syariah jika tidak dikembalikan ke MS dan QS
- A critical discourse developed in the 1990s in order to deconstruct (or 'to queer') sexuality and gender in the wake of gay identity politics, which had tended to rely on strategic essentialism. Opposed to gender essentialism, queer theorists see sexuality as a discursive social construction, fluid, plural, and continually negotiated rather than a natural, fixed, core identity.

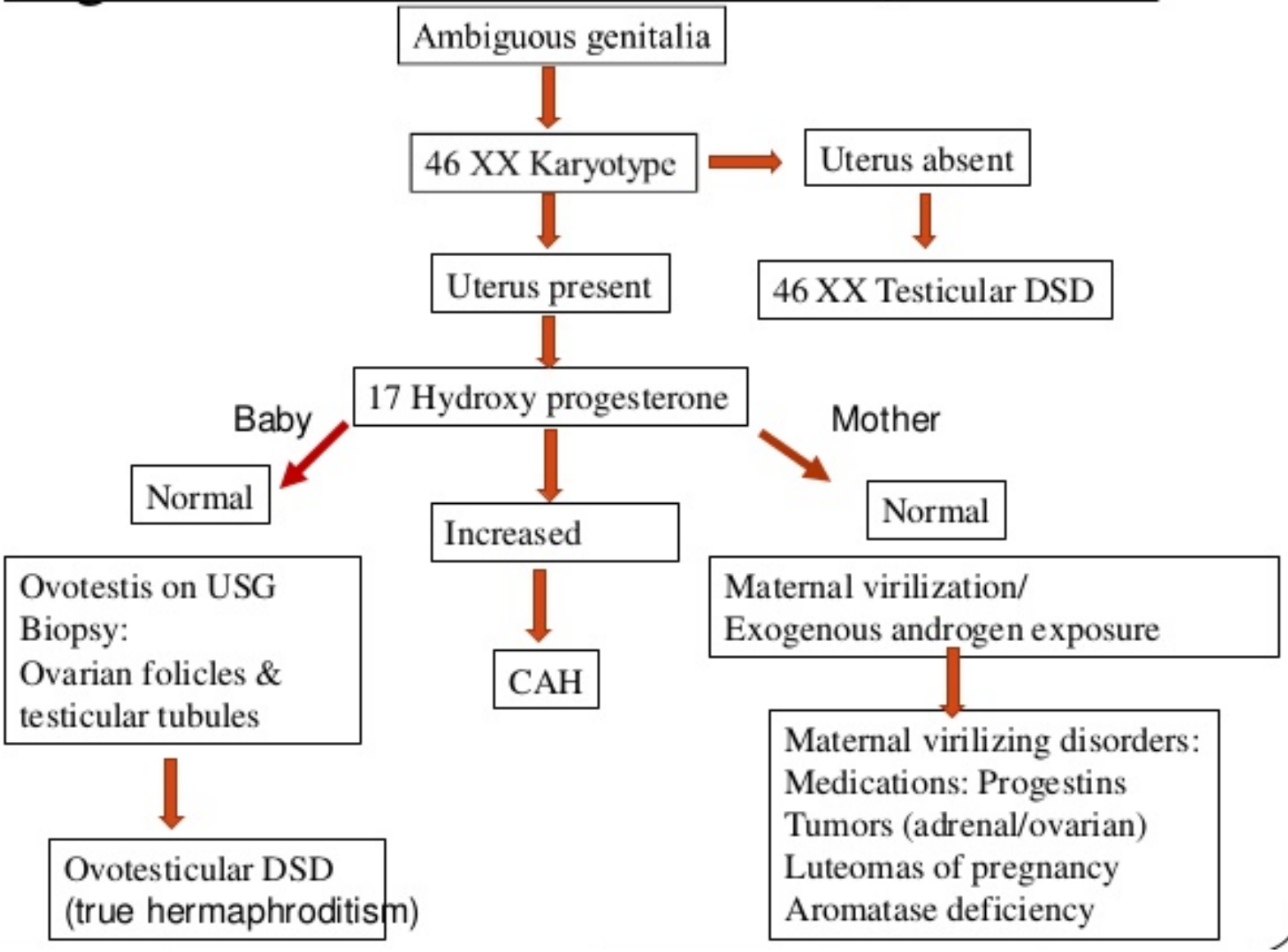
DIAGNOSIS?

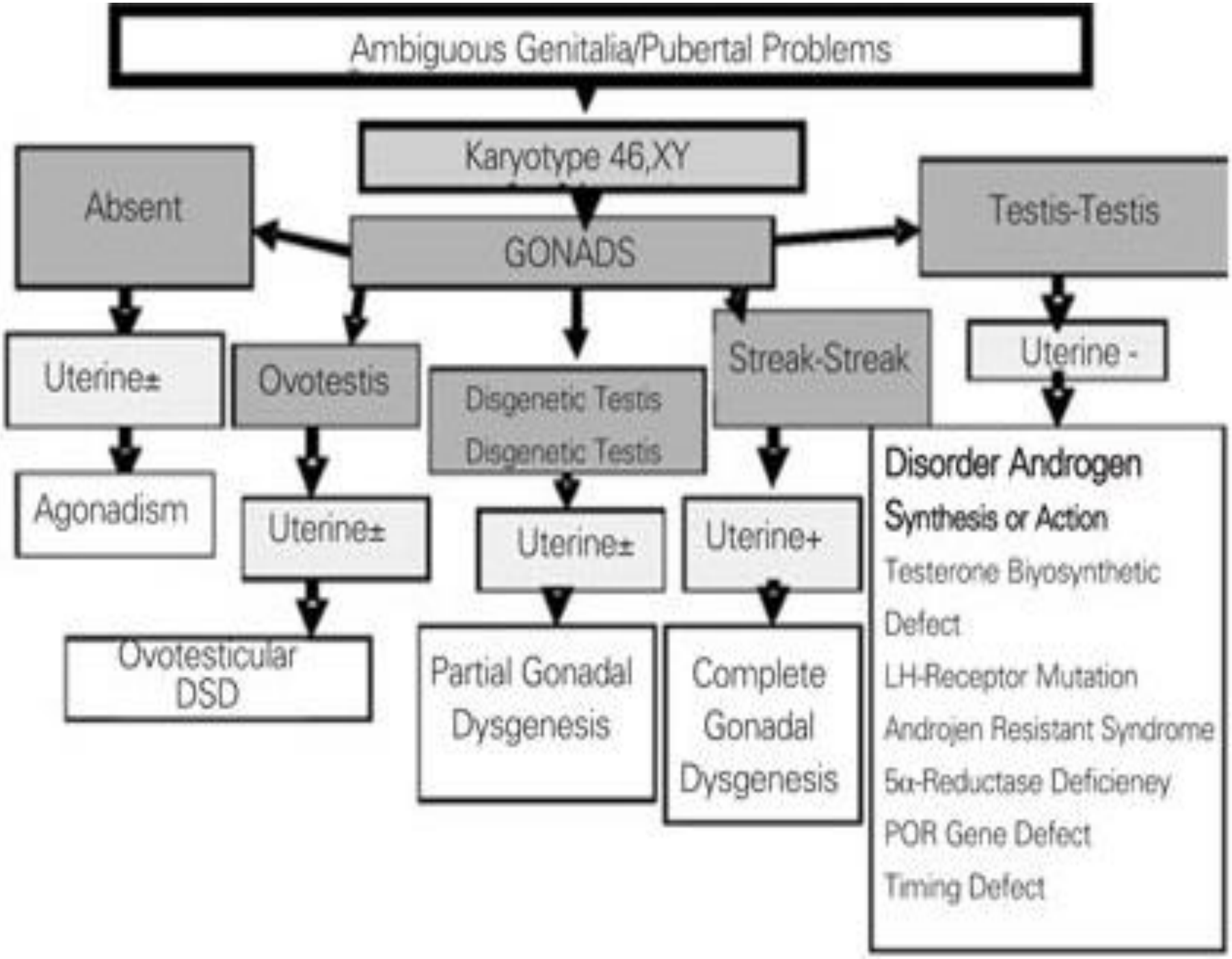
- The patient was of lateral variety in which testis was on right side and ovary on left side. The choice of rearing hermaphrodite as male or female sex is governed by phallus size
- In our patient penis was of adequate size thus plan in consultation with parents was made to rear him as a male. All female structures were thus removed. A repair of hypospadias will be performed in the next stage. Prosthesis can be placed in left hemiscrotum for psychological comfort.

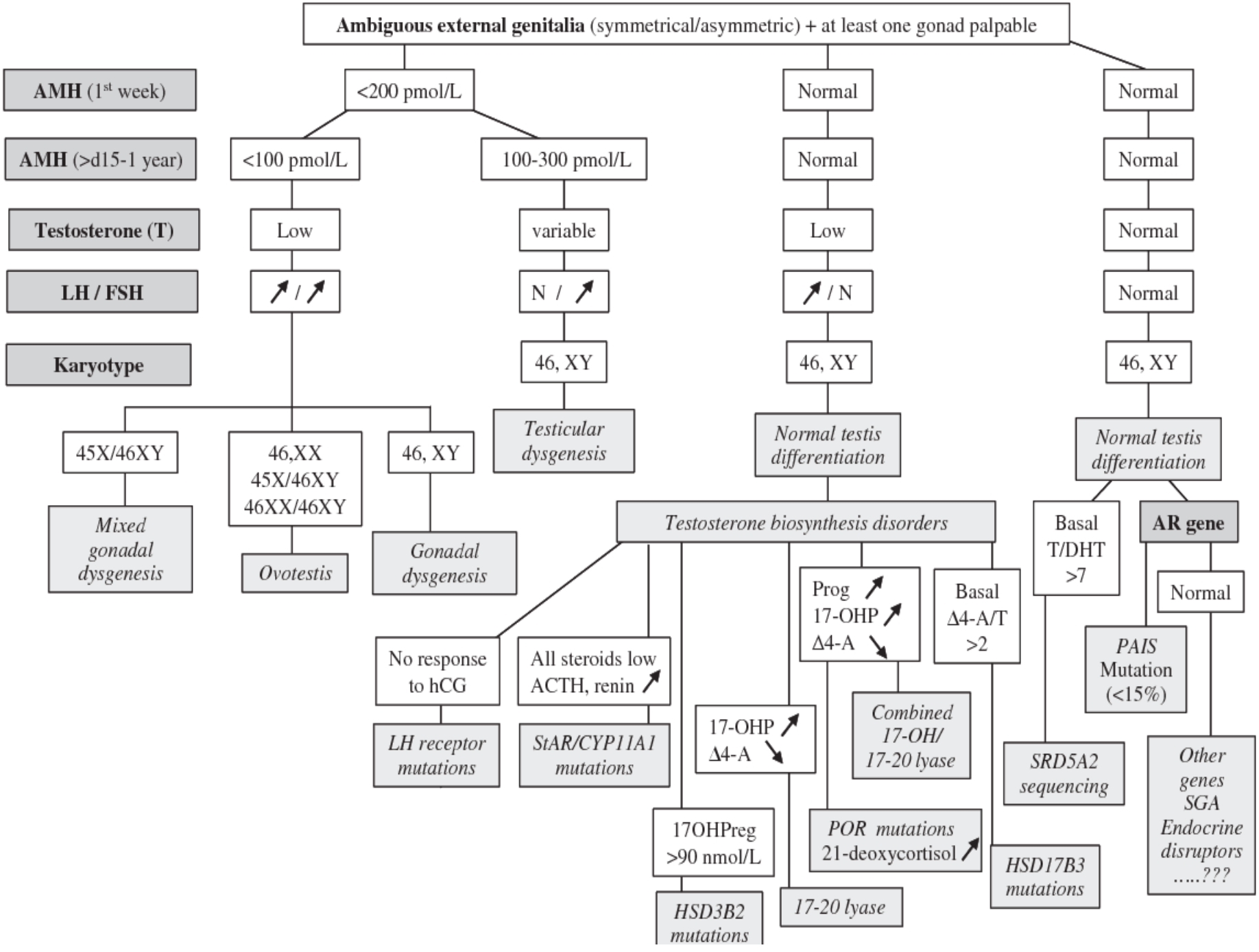
A

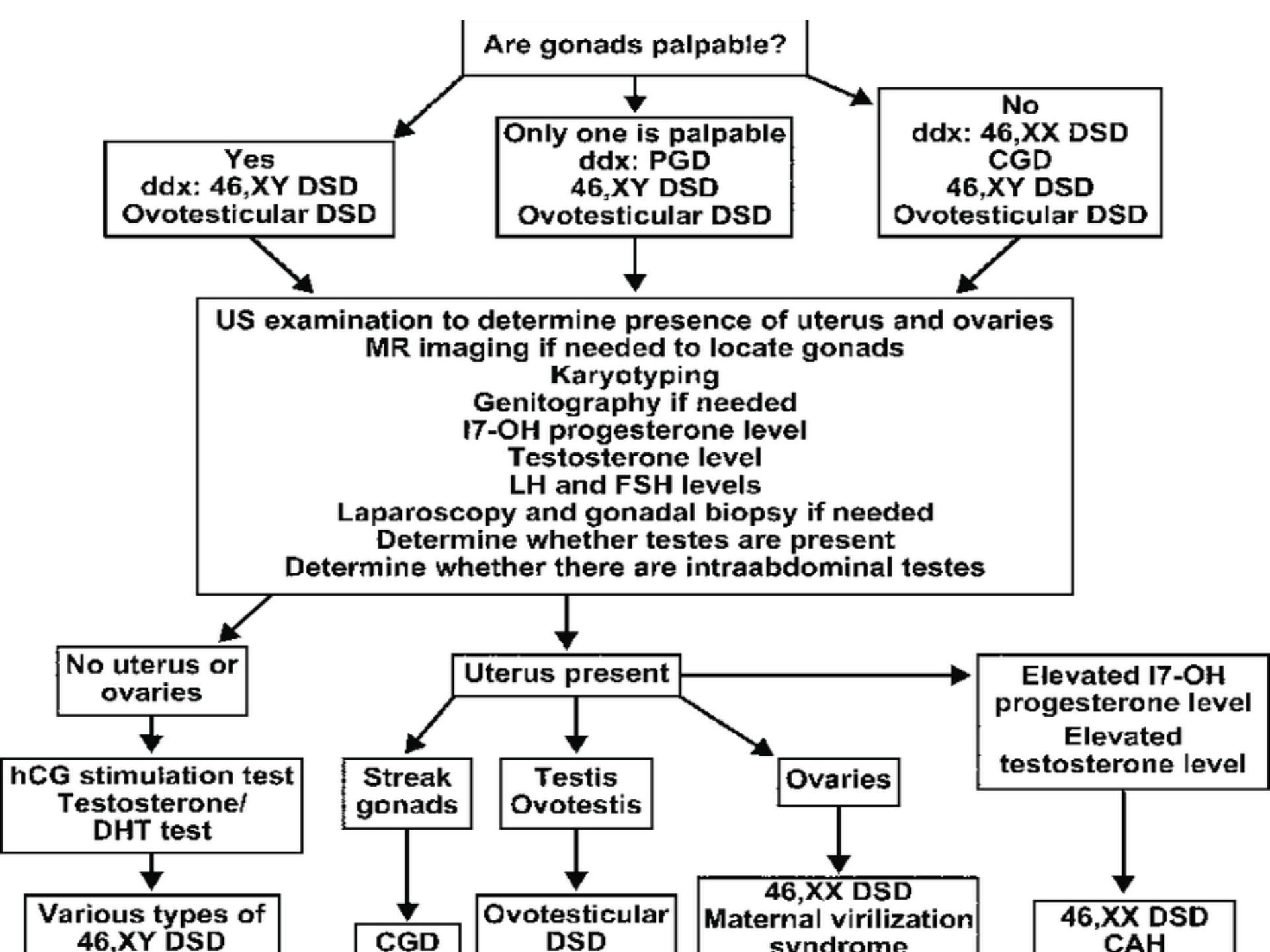

Ambiguous external genitalia
Karyotyping
46,XY
<3 months old
All age groups
Mutational analysis of SRD5A2 gene
Wild-type sequence
Mutation detected
5ARD ruled out
5ARD
N
USP
Characteristic findings
Defect in:
- 3β-HSD
- POR
- CYP17A1
- CYB5A
- 5AR
Baseline and post–ACTH-stimulated cortisol/baseline ACTH
Baseline and post–hCG-stimulated testosterone
Subnormal response/↑
N
N/↑
↓
Defect in:
- STAR
- CYP11A1
- SF1
AMH
AMH
N/↑
↓
N/↑
↓
Gonadal dysgenesis:
- Isolated: SRY, SF1, DAX1
- Syndromal: SOX9, WT1, ATRX, DHH, ARX, DMRT1, WNT4
- AIS
- PMDS (AMHR)
- Unknown factors
PMDS (AMH)
FSH/LH
↑
N/↓
Androstenedione
Hypogonadotropic hypogonadism
↑
↓
17β-HSD deficiency
- Leydig cell hypoplasia
- Partial gonadal dysgenesis

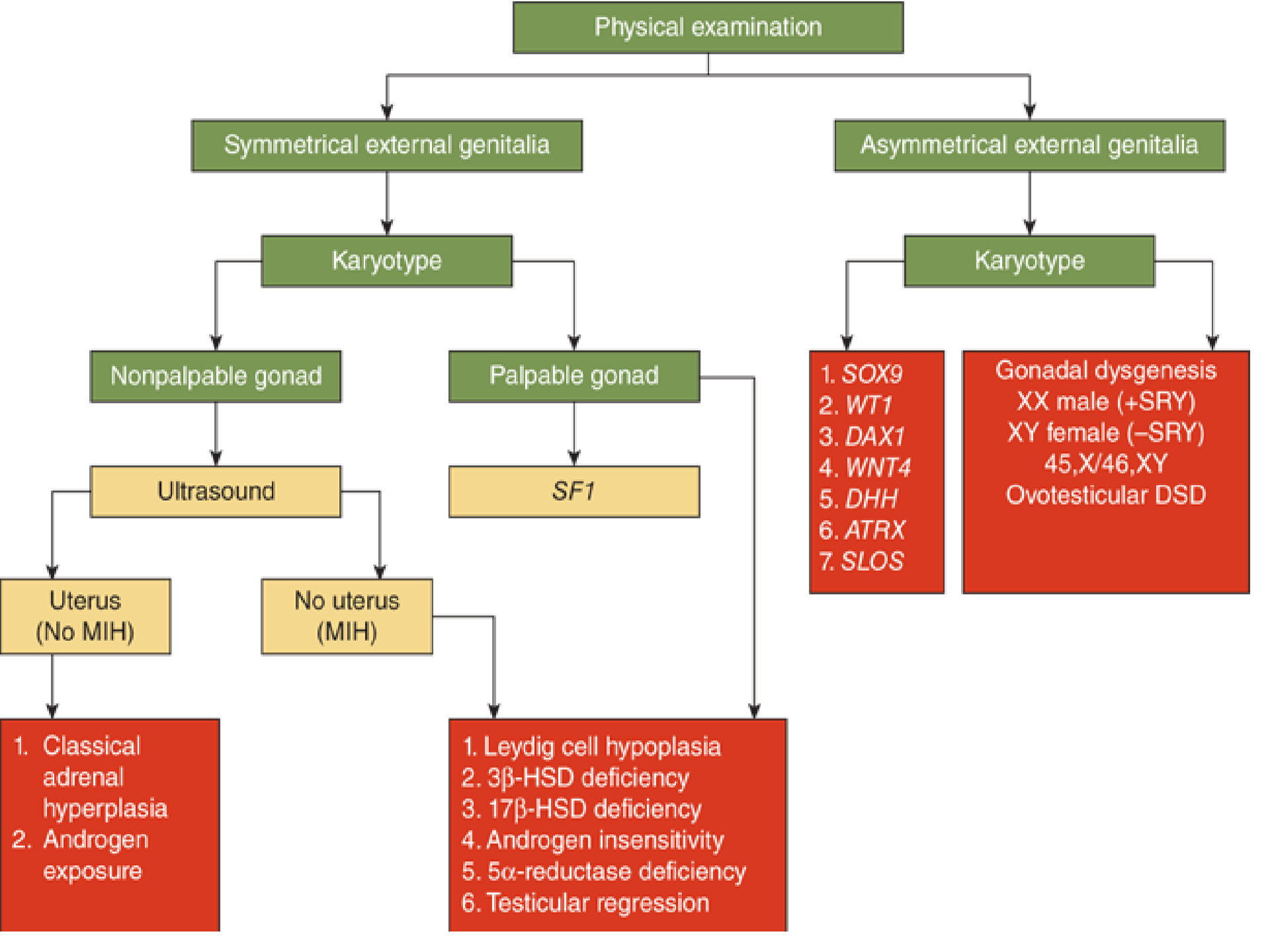
Algorithm for evaluation of 46,XX DSD




Ambiguous Genitalia/Pubertal Problems
Karyotype 46,XY
GONADS
Absent
Testis-Testis
Uterine±
Ovotestis
Disgenetic Testis
Disgenetic Testis
Streak-Streak
Uterine -
Agonadism
Uterine±
Uterine±
Uterine+
Disorder Androgen
Synthesis or Action
Testerone Biyosynthetic
Defect
LH-Receptor Mutation
Androjen Resistant Syndrome
5α-Reductase Deficieney
POR Gene Defect
Timing Defect
Ovotesticular
DSD
Partial Gonadal
Dysgenesis
Complete
Gonadal
Dysgenesis


Ambiguous external genitalia (symmetrical/asymmetric) + at least one gonad palpable
AMH (1st week)
AMH (>d15-1 year)
Testosterone (T)
LH / FSH
Karyotype
<200 pmol/L
Normal
Normal
<100 pmol/L
100-300 pmol/L
Normal
Normal
Low
variable
Low
Normal
↗ / ↗
N / ↗
↗ / N
Normal
46, XY
46, XY
46, XY
45X/46XY
46,XX
45X/46XY
46XX/46XY
46, XY
Testicular dysgenesis
Normal testis differentiation
Normal testis differentiation
Mixed gonadal dysgenesis
Ovotestis
Gonadal dysgenesis
Testosterone biosynthesis disorders
Basal T/DHT >7
AR gene
Prog ↗
17-OHP ↗
Δ4-A ↘
Basal Δ4-A/T >2
Normal
No response to hCG
All steroids low
ACTH, renin ↗
PAIS Mutation (<15%)
17-OHP ↗
Δ4-A ↘
Combined 17-OH/ 17-20 lyase
LH receptor mutations
StAR/CYP11A1 mutations
SRD5A2 sequencing
Other genes SGA Endocrine disruptors .....???
17OHPreg >90 nmol/L
POR mutations
21-deoxycortisol ↗
HSD17B3 mutations
HSD3B2 mutations
17-20 lyase


Are gonads palpable?
Yes
ddx: 46,XY DSD
Ovotesticular DSD
Only one is palpable
ddx: PGD
46,XY DSD
Ovotesticular DSD
No
ddx: 46,XX DSD
CGD
46,XY DSD
Ovotesticular DSD
US examination to determine presence of uterus and ovaries
MR imaging if needed to locate gonads
Karyotyping
Genitography if needed
I7-OH progesterone level
Testosterone level
LH and FSH levels
Laparoscopy and gonadal biopsy if needed
Determine whether testes are present
Determine whether there are intraabdominal testes
No uterus or ovaries
hCG stimulation test
Testosterone/ DHT test
Various types of 46,XY DSD
Uterus present
Streak gonads
CGD
Testis Ovotestis
Ovotesticular DSD
Ovaries
46,XX DSD
Maternal virilization syndrome
Elevated I7-OH progesterone level
Elevated testosterone level
46,XX DSD
CAH


Physical examination
Symmetrical external genitalia
Asymmetrical external genitalia
Karyotype
Karyotype
Nonpalpable gonad
Palpable gonad
1. SOX9
2. WT1
3. DAX1
4. WNT4
5. DHH
6. ATRX
7. SLOS
Gonadal dysgenesis
XX male (+SRY)
XY female (–SRY)
45,X/46,XY
Ovotesticular DSD
Ultrasound
SF1
Uterus
(No MIH)
No uterus
(MIH)
1. Classical adrenal hyperplasia
2. Androgen exposure
1. Leydig cell hypoplasia
2. 3β-HSD deficiency
3. 17β-HSD deficiency
4. Androgen insensitivity
5. 5α-reductase deficiency
6. Testicular regression

APA LANGKAHNYA?

- Clinical evaluation
- Diagnostic evaluation
- Surgical management
- Gender assignment in newborn infants
- Sex steroid replacement
- Psychosocial management
- Surgical Outcome

TINDAKAN BEDAH DAN PENGGUNAAN HORMON SEX

- Secara bioetika diperbolehkan tergantung pada tujuannya, adanya kewajiban, manfaat dan menjunjung otonomi

TABLE 1 Two studies of the sexuality in female patients with 21-OHD (reported a decade or more ago), in those with an early or late diagnosis, variable compliance with treatment, and more importantly, with different surgical procedures to repair the ambiguous genitalia

Criteria, %	USA [14] (80 patients)	Lyon [15] (25 patients)
Married	31	32
Single	69	68
Intercourse experience	62.5	60
No intercourse experience	37.5	40
Gynaecological examination and intercourse satisfactory	65	76
Stenosis of introitus	35	24
Active heterosexuality	57	60
Homosexual attraction	0	4
Active homosexuality	5	0
Fertility	20	16

Congenital Adrenal Hyperplasia

CAH is one the most common autosomoal recessive disorders resulting from a deficiency of one of the enzyme required for synthesis of cortiso in the adrenal cortex

More than 90% of cases are caused by 21 OHD

TABLE 1. Clinical features and electrolyte and hormonal data for thirteen Japanese female SW patients. Patients were classified based on Prader stages. All data were obtained before treatment. Normal age- and sex-adjusted reference values are shown in parentheses. Patients 7 and 8 were sisters. ND: Not determined

Patient	Prader stage	vaginal confluence	Na (mEq/l) (133-146)	K (mEq/l) (3.2-5.5)	Cl (mEq/l) (96-110)	PRA (ng/ml/hr) (< 26.2)	17OHP (ng/ml) (< 0.97)	ACTH (pg/ml) (13.8-37.8)	testosterone (ng/dl) (5.8-18.2)	DHEA-S (ng/ml) (43-1110)	17-KS (mg/day) (1.1-3.6)
1	1		128	5.5	102	100	150	125	111	ND	2.0
2	2		132	8.6	100	ND	270	1000	1430	1420	2.6
3	2		132	6.0	ND	96.5	311	820	790	2395	ND
4	2		127	6.9	ND	ND	198	1200	1370	ND	8.6
5	2		128	7.0	ND	226.8	211	814	344	1230	ND
6	3	low	132	7.1	107	8.2	626	800 <	ND	ND	ND
7	3	low	122	6.7	90	ND	586	ND	ND	ND	5.6
8	3	moderate	135	8.7	100	ND	174	770	ND	ND	ND
9	3	moderate	130	7.8	ND	ND	120 <	ND	ND	ND	ND
10	3	high	126	7.8	ND	ND	221	1600	ND	ND	ND
11	4	high	127	8.8	99	> 20	200	464	ND	ND	ND
12	4	modelate	135	5.2	105	> 20	608	520	ND	ND	1.6
13	4	low	135	5.4	104	25.2	119	529	125	517	ND

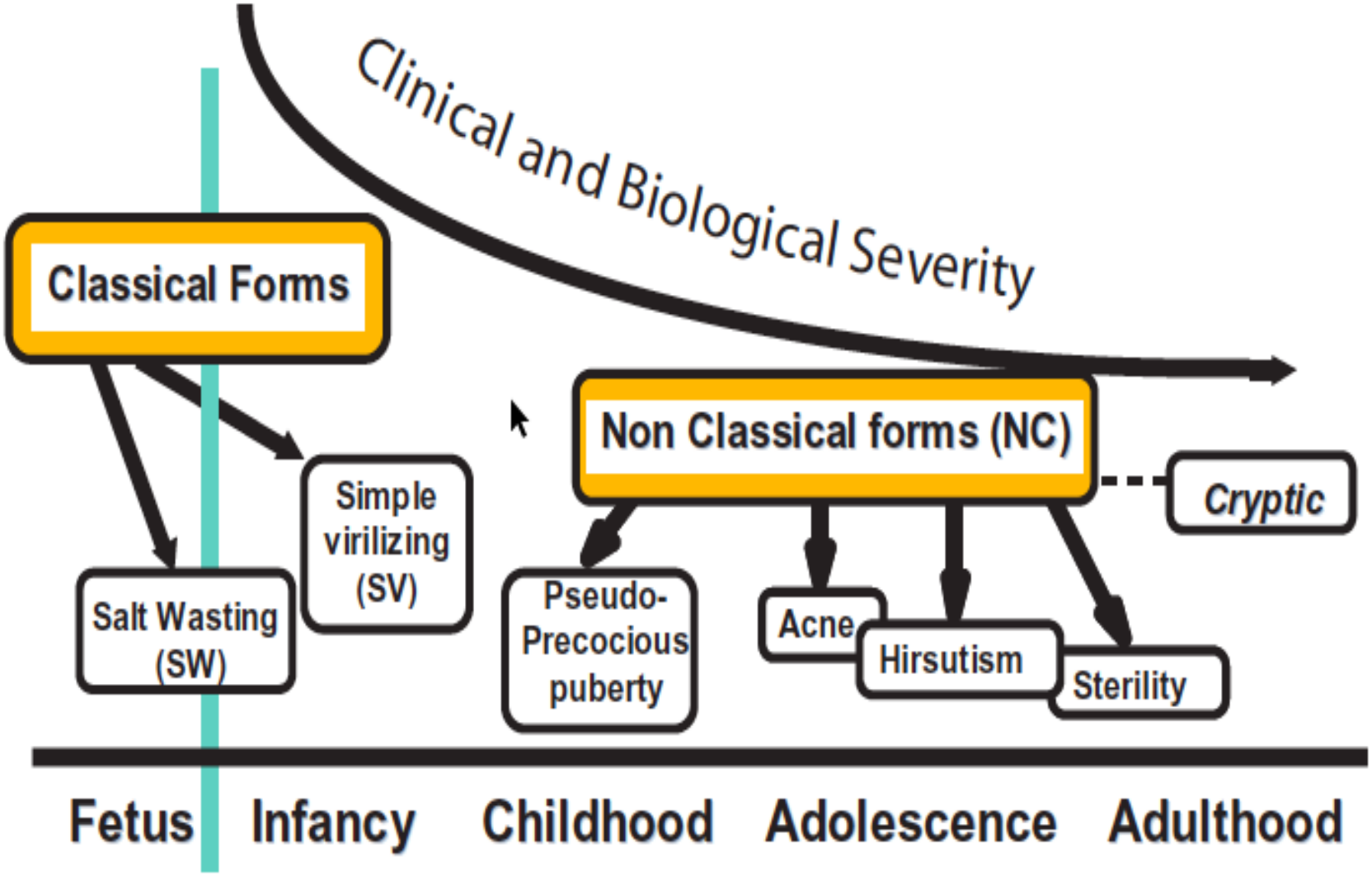
Patient	Present age	Age at visiting the clinic	Reasons of visiting the clinic	Prader stage
1	3y3mo	8 d	mass screening	1
2	14y5mo	1 d	ambiguous genitalia	2
3	6y8mo	1 d	ambiguous genitalia	2
4	14y0mo	0 d	ambiguous genitalia	2
5	1y8mo	1 d	ambiguous genitalia	2
6	25y9mo	7 d	ambiguous genitalia	3
7	28y0mo	1 d	ambiguous genitalia	3
8	24y3mo	1 d	ambiguous genitalia	3
9	5y2mo	1 d	ambiguous genitalia	3
10	8y10mo	9 d	ambiguous genitalia	3
11	4y6mo	8 d	mass screening	4
12	23y6mo	7 d	ambiguous genitalia	4
13	6y0mo	1 d	ambiguous genitalia	4

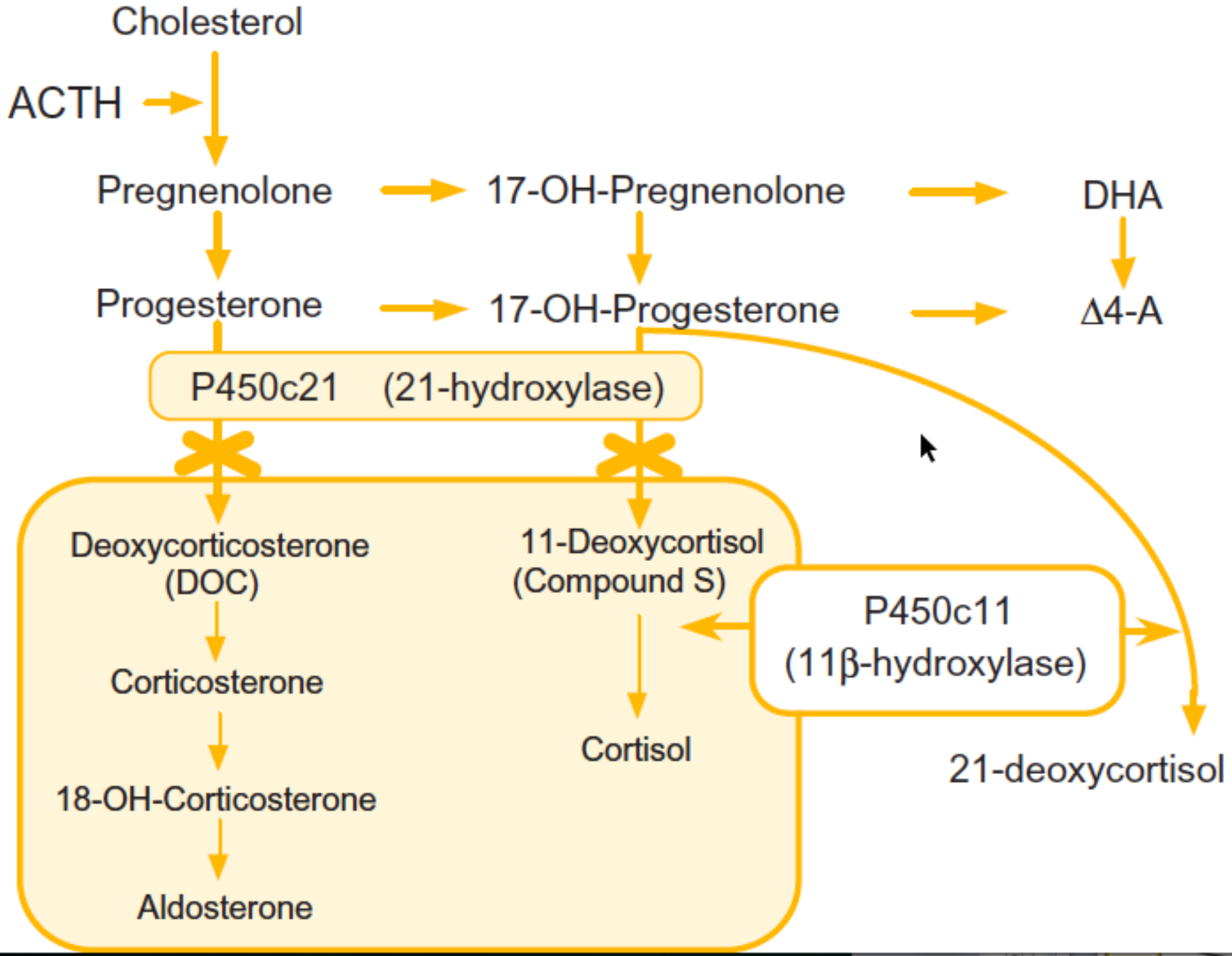
Table 1. Mutations in Genes Involved in Sex Determination and Development and Associated with Intersex Anomalies.

Gene (Locus)	Protein and Proposed Function	Mutant Phenotype
WT1 (11p13)	Transcription factor	Frasier syndrome, Denys–Drash syndrome with Wilms' tumor
SF-1 (9q33)	Transcription factor, nuclear receptor	Gonadal and adrenal dysgenesis
SOX9 (17q24)	High-mobility-group transcription factor	Campomelic dysplasia, male gonadal dysgenesis or XY sex reversal
DAX1 (Xp21.3)	Transcriptional regulator, nuclear-receptor protein	Gonadal dysgenesis, congenital adrenal hypoplasia
SRY (Yp11)	High-mobility-group transcription factor	Gonadal dysgenesis
MIS, or *AMH*, type II receptor (12q12–13)	Serine threonine kinase receptor	Persistent müllerian duct syndrome
MIS, or *AMH* (19p13)	Secreted protein, causes regression of fetal müllerian duct; Leydig-cell inhibitor	Persistent müllerian duct syndrome
AR (Xq11–12)	Androgen receptor, a ligand transcription factor	Male pseudohermaphroditism, complete or partial androgen insensitivity syndrome
HSD17B3 (9q22)	17β-Hydroxysteroid dehydrogenase, 17-ketosteroid reductase 3	Male pseudohermaphroditism
SRD5A2 (5p15)	5α-Reductase type 2	Male pseudohermaphroditism*
CYP17 (10q24–25)	17-Hydroxylase: 20–22 lyase	Male pseudohermaphroditism
CYP21 (6q21.3)	21-Hydroxylase	Congenital adrenal hyperplasia, female pseudohermaphroditism
HSD3B2 (1p13.1)	3β-Hydroxysteroid dehydrogenase type II	Congenital adrenal hyperplasia
CYP11B1 (8q24)	11β-Hydroxylase	Congenital adrenal hyperplasia
StAR (8p11.2)	Steroidogenic acute regulatory protein	Congenital lipoid adrenal hyperplasia

* Virilization may occur at puberty.

FIG. 4. Spectrum of the clinical forms of 21-OHD. The classic forms (virilizing ones) have a fetal onset while the non-classic have a variable timing of clinical onset long after birth.




Cholesterol
ACTH
Pregnenolone
17-OH-Pregnenolone
DHA
Progesterone
17-OH-Progesterone
Δ4-A
P450c21 (21-hydroxylase)
Deoxycorticosterone (DOC)
11-Deoxycortisol (Compound S)
P450c11 (11β-hydroxylase)
Corticosterone
Cortisol
21-deoxycortisol
18-OH-Corticosterone
Aldosterone

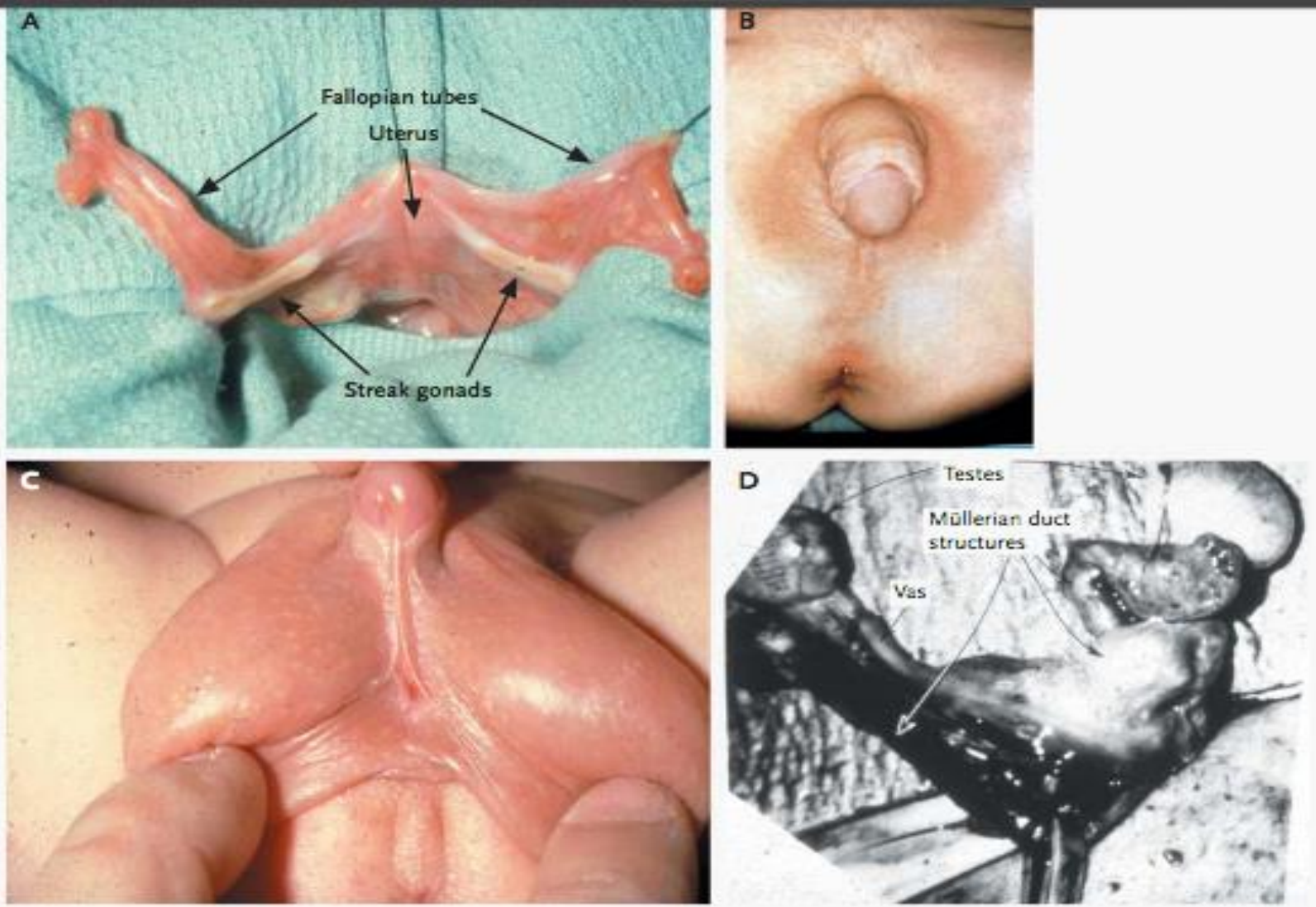


Figure 1. Clinical Examples of Intersex Abnormalities.

Panel A shows the internal structures of a 46,XY patient with pure gonadal dysgenesis. There are bilateral streak gonads with retained müllerian structures, fallopian tubes, and a midline uterus. Panel B shows severe clitoral hypertrophy caused by masculinization of the external genitalia of a 46,XX patient with female pseudohermaphroditism caused by congenital adrenal hyperplasia. Panel C shows incomplete masculinization of the external genitalia of a 46,XY patient with male pseudohermaphroditism. There is a microphallus with perineoscrotal hypospadias and bifid and prepenile scrota. Panel D shows internal genitalia of a 46,XY patient with persistent müllerian duct syndrome. There are müllerian structures (i.e., fallopian tubes and uterus) as well as wolffian structures (i.e., the vas deferens and epididymis). Panel B

PENUTUP

- Intersex merupakan murni kelainan bawaan yang sudah ditetapkan konsensus cara penegakan diagnosisnya dan penanganannya
- Penanganan medis dilakukan untuk mengembalikan fertilitas, fungsi reproduksi/prokreasi kembali normal dengan tetap menjaga rekreasi errotic
- Kesesuain fenotip dan genotip meliputi cromosom, hormon, organ genital interna dan eksternal, dan gonad sex
- Tindakan pembedahan sangat diperlukan dengan disertai pemberian terapi hormonal
- Prinsip bioetik otonomi, beneficence, non maleficence dan justice harus diutamaka dalam melakukan tindakan yang tepat
- Bioetik islam terkait dengan isu intersex adalah sesuai dengan MQ dan QS SEHINGGA MENGUBAH KELAMIN DIPERBOLEHKAN SESUAI TUJUAN MS