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South African Society for Ultrasound in Obstetrics and Gynaecology.

CLINICAL GUIDELINE

This document is intended to guide clinical care without changing the responsibility of the health care team or the patient. It never replaces clinical judgment and individualized care.

Developed by SASUOG and endorsed by SASOG as part of the BetterGYN® programme

SASUOG POSITION STATEMENT ON THE LATE TERMINATION OF PREGNANCY FOR FETAL ANOMALIES (TOPFA)

*The aim of this position statement is to contribute towards **balanced, transparent, consistent (over time) and uniform (between practices) decision making** when abnormal fetal development is detected after 20 weeks, thereby improving equitable access to late TOPFA.*

Preamble:

1. Access to early prenatal diagnosis has improved but due to continuing problems of late presentation and late referral, the number of women in whom fetal anomalies are diagnosed late in pregnancy has increased.
2. A guideline is needed indicating conditions for which clinicians are expected to offer TOPFA and others for which late TOPFA is not considered appropriate since the wording of clause c) ii) of the South African Choice on Termination of Pregnancy (CTOP) Act of 1996 (TOP after 20 weeks is allowed "if continuation of the pregnancy would result in a severe malformation of the fetus") is vague and the meaning of "severe" is untested in the judicial system and open to wide interpretation.
3. In late gestation there is a distinct difference between the process of ending the pregnancy (addressed in the CTOP Act) and the pregnancy resulting in a non-surviving infant (not addressed in this Act as it only deals with issues related to the "Separation and expulsion, by medical or surgical means, of the contents of the uterus of a pregnant woman"). In late gestation, non-survival may require a feticide procedure.

4. The basis for this document is the Tygerberg Hospital (TBH) Policy document on late TOPFA. Details about the development of this document can be found in Addendum 1. The document was approved by the TBH Clinical Ethics Committee and hospital management in 2015 and has been consistently implemented since. Heads of fetal medicine units at several other academic departments in South Africa have asked permission to use the policy document in their own setting and they have supported the principles represented in it.

Fundamental principles of this position statement:

1. The health care sector must be **committed to continue building services that facilitate access to early prenatal diagnosis** in an effort to reduce the number of cases where late TOPFA needs to be considered.
2. A fair, consistent and transparent policy can best be achieved by **relying on ethical principles and professional integrity**
3. While the **presumption must always be in favour of life,** this inherent respect for life, including life before birth, is not seen as absolute but as **potentially rebuttable, but then for grave reasons only**. To limit both an unduly liberal or unreasonably conservative approach, reasons for late TOPFA should be defined in sufficient detail, taking the interests of the mother as well as the prospective child into consideration.
4. The reasons for rebuttal must be increasingly severe as the pregnancy advances, supporting the principle of **gradualism** in this respect for life.
5. In cases of uncertainty or disagreement or when a late TOPFA is declined by the team despite the parents' request, a **review or appeal process should be available.**
6. Clause c) ii) of the SA CTOP Act is understood to mean the following:
    a. The term "termination of pregnancy" is reserved for interventions that result in non-surviving offspring. For TOPFA at a potentially viable gestation this will usually require a feticide procedure, unless the condition is uniformly and unequivocally lethal in the very short term. It is understood that the principles that regulate TOPFA **invariably apply to feticide** procedures, irrespective of whether elective delivery follows feticide or not.
    b. "Severe malformation" is understood to include **any abnormality of body or function that will have a severe impact on the quality of life of the infant and child** rather than only "malformations" which literally would exclude congenital infections, many genetic conditions, mental disability etc.
    c. **"Severe" needs to be ethically justified** by medical professionals who act with integrity and always aspire to "do the right thing". A very liberal interpretation of the law, which would allow eugenic practices with excessive use of TOPFA to avoid any risk of a birth defect, is therefore not supported.
7. Decisions regarding late TOPFA are best made by a **multidisciplinary team of knowledgeable professionals.** While absolute certainty about diagnosis and outcome is virtually impossible for most conditions, the clinical team needs to undertake meticulous assessment of the cases with use of the most recent literature and wide consultation of relevant clinicians and allied health care professionals as deemed necessary, to obtain the **best possible opinion** about the real prognosis of the anomaly in an individual case.

8. The prognosis needs to be determined within the local context and according to current (and prospective) realistically available care and treatment for this specific infant and taking all the specifics of the case into account, including the impact on outcome by a combination of different anomalies. As a reference, it needs to be considered what neonatologists and paediatric subspecialists would regard as appropriate care in case such a child was born alive. The mother's social context may affect the likely outcome of a condition and needs to be taken into account in the decision-making.

9. For anomalies where a spectrum of outcomes is possible but where the severity of the outcome in the individual case cannot be determined accurately, the decision regarding the need to offer late TOP will be based on the **average or most common expected outcome** and **not on the rare, worst (or best) case** scenario. It may not be possible to define strict criteria regarding which probability of good (or poor) outcome would justify the offer of late TOP but, as a general principle, for conditions with a similar average outcome, a request for late TOP may be declined if a significant minority is expected to have a good or very good outcome while it may be granted if the best expected outcome still encompasses significant impairment or if the degree of disability or suffering in the worst case scenario is considerable.

10. A differential approach is used according to the gestation at detection, employing somewhat more liberal criteria for diagnoses made in the first few weeks after 20 weeks and more strict criteria for conditions that are diagnosed at a stage when the fetus would already be capable of living independently from the mother's body. This is based on the following realities:

    a. before viability, the fetus has no prospect of becoming a person but for the continued support of the mother's body

    b. after viability, the fetus could already be a person but for the act of actively killing it

    c. the vast majority of fetal anomalies in this country are diagnosed after 20 weeks

    d. for many of these anomalies, TOP after 20 weeks was already legally offered in all South African academic institutions under the 1975 Abortion and Sterilisation Act, up to a gestation where a live birth became likely

    e. the 1996 CTOP Act was intended to be more liberal than the 1975 Abortion Act

    f. the concept of gradualism is embedded in the current CTOP Act which clearly implies that the best interests of the prospective child increasingly outweigh maternal choice as fetal development progresses

    g. most countries where late TOPFA is legal use a higher cut-off than 20 weeks to differentiate severity criteria

    h. the chance of a live birth after induced labour for TOPFA is very gestational age dependent but similar in all settings. Live birth is extremely rare before 22 weeks but in contrast to this, 60% of fetuses are born alive after labour at 24 weeks, and less than a fifth of these die in the first few hours of life

    i. 24 weeks can therefore serve as a pragmatic definition of the gestational age at which more restrictive criteria for TOPFA are to be employed

Practical guideline based on the fundamental principles

1. To translate these principles into a practical and pragmatic guide for the multidisciplinary clinical team, it was concluded that **specific fetal conditions can usually be placed in one of 4 groups**, which are distinguishable on ethical grounds:

    a. Group1: Conditions that always qualify for late TOPFA, irrespective of gestation and presentation.

    b. Group2: Conditions that only qualify for late TOPFA in individual cases that meet certain severity criteria (severity in the individual can be determined with acceptable accuracy).

    c. Group 3: Conditions that will generally not be considered for late TOPFA at advanced gestation.

    d. Group 4: Conditions that do not meet strict criteria for group 1 and 2 but where individual characteristics of the anomaly or circumstances of the family may constitute significant aggravating factors warranting further assessment for an individualised decision.

2. The **ethical principles and criteria that define the specific groups** are described in sufficient detail to inform decisions in the future and a list of **example anomalies** typical of each category is provided for illustrative purposes (Addendum 2). These lists are not exhaustive and may change over time as and when new evidence becomes available.

GROUP 1 AND 2 CONDITIONS – TOPFA TO BE OFFERED AT ANY GESTATION

1. There must be **(near) certainty of diagnosis and (near) certainty of outcome** and the expected outcome (with currently and realistically available standard of care and taking all the specifics of the case into account, including the impact on outcome by a combination of different anomalies) is one (or more) of the following:

    a. Early death in spite of currently and realistically available standard of care. This refers to conditions where active intervention is regarded as futile since it often only prolongs suffering with little or no benefit in terms of long-term survival (suggested definition: > 90% death in infancy).

    b. Profound and irreversible deficit in developmental capacity, with one or more disabilities (intellectual, visual, hearing, physical), resulting in inability to achieve a reasonable level of self-awareness or reasonable level of functioning within society or inability to develop meaningful interpersonal relationships. This refers to conditions for which active intervention is regarded as futile since it often only prolongs suffering with little or no benefit in terms of developing the capacity to experience human life in a meaningful way.

    c. Unbearable pain and suffering on the part of the child, in order to survive, with at best a very poor quality of life being anticipated.

    d. Unreasonable burden of care on the part of the parents or society when an unreasonable amount of medical care will be needed to ensure a reasonable quality of life, which is deemed unfeasible within the current context of available services and resources (this can be specific to the individual family unit and could include conditions with fully dependent ultimate performance). The assessment of whether the burden of care is deemed

unreasonable is determined by a multidisciplinary team including a subspecialist knowledgeable in the specific anomaly/anomalies and, when needed, a social worker.

2. In summary, infants in this group **will always, at best, be severely compromised**.

3. Neonatologists and paediatric subspecialists will generally only offer palliation and comfort care or have a low threshold for withholding or withdrawing active intervention.

GROUP 3 CONDITIONS – TOPFA IS NOT OFFERED AFTER VIABILITY

1. Conditions in group 3 have one or more of the following characteristics:

   a. **Long terms survival** is anticipated.

   b. Almost invariably associated with the **capacity to meaningfully experience life and to fulfil a meaningful role in human society, without unbearable suffering.**

   c. **Paediatricians generally offer standard of care** once the child is born, to optimise the outcome and maximise the potential to a fulfilling life.

   d. Considered trivial or **readily correctable** or have a low risk for abnormal outcome in the long term.

   e. May be associated with long term disability but where the **disability is regarded as being manageable**, even if this involves a significant burden on the part of the child, the parents and/or society and even if the ultimate outcome involves assisted performance or the use of aids to obtain the best possible result. The fact that the child may have some form of disability or may be disadvantaged to some extent does not exclude conditions from this group.

2. It is reiterated that the anticipated favourable outcome which defines this group needs to be seen **in the local context according to current (and prospective) realistically available treatment for this specific infant** (at this specific point in time and taking all the details of the case into account) and needs to be based on evidence from the literature as being the **outcome in the majority** of cases, even though it is understood that the outcome may be better in some individuals and worse in others. If, on paediatric reassessment after delivery, the prognosis is deemed unexpectedly severe in a specific individual, paediatric care will be amended accordingly, as is current practice. Several of these anomalies **may require ongoing monitoring and review**, possibly with additional imaging modalities later in gestation to detect any deterioration over time or any new features that may alter the decision regarding late TOPFA.

3. The criteria for group 3 conditions are grounded on a **strong beneficence-based obligation** that exists towards the fetuses in advanced stages of development, not only on the part of medical professionals but also on the part of the parents. Once a pregnancy has progressed so far that the fetus could potentially survive independently from the mother's body, the fetal interests may legitimately take preference over maternal choice. While maternal choice outweighs the fetal interests for the severe abnormalities of group 1 and 2, the interests of the prospective child take preference over maternal choice for the non-severe group 3 conditions. For non-severe conditions, a mother is reasonably expected to agree to interventions

that are reliably expected to benefit her unborn child, including continuing with the pregnancy.

4. Given the lack of any ethical or medical distinction between fetuses of 21 and 19 weeks while there are massive medical and ethical differences between fetuses of 21 and 41 weeks, TOPFA can be offered in the first few weeks after 20 weeks for some anomalies in group 3 based on the arguments explained under point 10 of the fundamental principles.

    a. The anomalies should at least be compatible with the CTOP Act prior to 20 weeks ("there is a **substantial risk** that the fetus would suffer from a severe physical or mental abnormality") while not meeting the level of certainty for TOPFA after 20 weeks as currently described in the CTOP Act ("continuation of the pregnancy **will result** in a severe malformation").

    b. For these anomalies there should be a reasonably high likelihood of a significantly limited quality of life, a significantly limited level of functioning, considerable suffering, or considerable burden of care in the long term.

    c. Twenty-four completed weeks is the suggested gestational age after which TOP for these anomalies is no longer offered but this may vary (according to fetal size, the severity of the anomaly, the likelihood of a live birth and the acceptability thereof not only to the parents but also the labour ward and paediatric team, the anticipated suffering in case of a live birth and the potential survival etc.).

GROUP 4

1. **Conditions that do not meet strict criteria** for group 1 and 2 but where the specific case is of exceptional severity or where individual circumstances of the pregnancy or the family are of such an **aggravating** nature that they would significantly impact on the ultimate outcome for the infant.

2. Examples of such circumstances could be the inevitability of severe prematurity, the inability of the family unit to access essential medical services, the unfavourable combination of different anomalies etc.

3. The main criteria in this deliberation, to justify a more individualised decision would be the **expected quality of life of the specific infant, given the health care it would realistically receive in the specific circumstances**.

4. This is a difficult group and extensive consultation, involving an in-depth assessment of the psychosocial circumstances, is indicated. If no consensus can be reached, the case may warrant review by the Institutional Ethics Committee.

ADDENDUM 1

The development of the Tygerberg Hospital Policy Document on late TOPFA

This policy document was drafted by clinicians from multiple disciplines involved in the care of women whose fetuses are diagnosed with abnormal development (and often their children, once delivered) and compiled after review of the literature, attempting to develop an understanding of the different views and opinions as well as the ethical principles needed in guiding decision making in this difficult area.

Development of the Tygerberg Hospital (TBH) policy document took place over more than two years and started with journal club sessions with Fetal Medicine subspecialty fellows, clinical genetics and obstetric registrars as well as a series of open discussions on a case-base within the clinical team, always with a wide input of relevant subspecialists.

In October 2012, the TBH Clinical Ethics Committee (CEC) requested the clinical team to produce an official written policy document to guide practice in this matter. For this purpose, a series of focus group discussions were arranged during 2013, with representation from fetal medicine, obstetrics, medical genetics, genetic counselling, paediatrics, ethics and hospital management. The discussions were transcribed and broad themes, agreements and proposals were summarized. The summary document was re-discussed in the focus group until broad consensus was reached regarding the principles of the proposed approach. Further (technical) refinements to the document were made by the clinical multidisciplinary team.

The draft document was presented to several people and professional groups for additional input and comments: Prof A Skelton (Professor of Law and Director of the Centre for Child Law at the University of Pretoria and knowledgeable on the conflict between fetal interests and reproductive health rights of women), Prof W Landman (Extraordinary Professor in the Centre for Applied Ethics and Head of Unit for Bioethics of Stellenbosch University), TBH neonatology consultants, the TBH O&G consultant and registrar body, members of TBH Nursing Management and the TBH Hospital board with representatives from the community and hospital administration. The document received wide support and no major objections were presented by any of the groups invited to provide feedback.

The document was approved by the Tygerberg Hospital Clinical Ethics Committee and hospital management in 2015.

ADDENDUM 2:

Illustrative examples of the different groups

Group 1: Late TOP is always offered, irrespective of gestation and presentation, as the outcome is almost uniformly dismal.

- Trisomy 13, 18, triploidy, rare trisomies
- Anencephaly, exencephaly, iniencephaly, craniorachischisis
- Alobar holoprosencephaly, hydranencephaly
- Otocephaly, lethal skeletal dysplasias, sirenomelia
- Acardia, ectopia cordis, hypoplastic left heart syndrome
- Limb-body-wall defect
- Bilateral renal agenesis or non-functional dysplastic kidneys
- Fetal akinesia sequence
- Severe inborn errors of metabolism, e.g., Tay-Sachs, Lesh-Nyan
- Severe genetic conditions, e.g., spinal muscular atrophy type 1, Meckel-Gruber syndrome

Group 2: Conditions with a spectrum of presentations that ONLY qualify for late TOP if certain severity criteria are met (severity in individual cases can be determined with acceptable accuracy).

- Severe unbalanced translocations
- Cytomegalovirus infection: with severe abnormalities on ultrasound
- Conjoined twins: with joined vital organs
- Non-immune hydrops (2 cavities with anasarca): with no treatable cause (incl.T21, 45,X and severe cardiac defects)
- Classic Dandy-Walker malformation: with severely hypoplastic cerebellum
- Microencephaly: < -3SD with evidence of abnormal parenchyma
- Severe schizencephaly
- Encephalocele containing brain: with abnormal intracranial CNS images
- Lower urinary tract obstruction: with severe oligohydramnios (not otherwise explained), abnormal renal texture (severely echogenic cortex or cortical cysts), abnormal urinary biochemistry or pulmonary hypoplasia
- Congenital diaphragmatic hernia: with severe pulmonary hypoplasia
- Severe cardiac defects: operability unlikely or multiple surgeries required but still with very limited life expectancy
- Severe osteogenesis imperfecta: with multiple fractures

- High open spina bifida: last intact vertebra L3 or higher
- Multiple malformations involving vital organs
- Exposure to medical teratogens such as retinoic acid derivatives, warfarin, methotrexate, efavirenz, dolutegravir: with severe abnormalities of essential organs (e.g., CNS, cardiac) on ultrasound

Group 3: Late TOP is NOT offered after 24 weeks

- Uncomplicated sex chromosome abnormalities, Trisomy 21, Di George, Fragile X syndrome
- Genetic conditions like Duchenne's muscular dystrophy, haemophilia A and B, sickle cell disease, cystic fibrosis
- Lower urinary tract obstruction with normal renal texture, amniotic fluid, lung volumes and urinary biochemistry
- **Apparently isolated** (after appropriate special investigations and review) moderate ventriculomegaly, hypoplasia of the vermis, complete or partial absence or hypoplasia of the corpus callosum, cleft lip and palate, micrognathia, talipes, absent limb, abnormal limb
- Major cardiac anomalies that are operable with a good surgical result and a favourable long-term outcome
- Confirmed fetal CMV infection without any ultrasound signs of damage
- Uncomplicated exomphalos, gastroschisis
- Achondroplasia
- Low open spina bifida, hemivertebrae

Group 3: Late TOP is offered after 20 weeks but not after 24 weeks

- Turner's syndrome, Trisomy 21, Di George, Male fragile X syndrome, Duchenne's muscular dystrophy, cystic fibrosis, sickle cell disease
- Moderate ventriculomegaly, partial aplasia of the vermis, complete or partial absence or hypoplasia of the corpus callosum
- Cardiac anomalies for which extensive and repeated surgery is needed with only reasonable medium-term outcome
- Confirmed symptomatic fetal CMV infection
- Giant exomphalos
- Severe micrognathia
- Low open spina bifida

RECOMMENDED READING

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