

Summary of Clinical Recommendations:

Approved at the Danish guideline-meeting, January 22nd, 2015
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The guideline is based on the recommendations from DFMS and FMF for the second trimester anatomy scan in which checking for SUA is mandatory. In Denmark, midwives routinely examine the umbilical cord after delivery for the number of vessels.

Clinical Recommendations

*Strength
Oxford system*

It is recommended to check for SUA in the second trimester according to the DFMS guideline.	√
It is recommended to evaluate aa. umbilicalis at the bladder level and possibly in the free-floating umbilical cord.	√
It is recommended to investigate the number of vessels in the cord after delivery.	√
The second-trimester anatomy scan should be conducted by health care professionals who are educated and certified according to FMF standards. If SUA is detected, extra attention is warranted to detect possible associated malformations.	√
When SUA is detected, a thorough fetal heart scan is warranted. At a minimum, a 4-chamber view, OFT and 3-vessel view should be performed in sufficient detail.	C
If the anatomy scan is not sufficiently performed (not all structures seen in detail), or if associated malformations (niSUA) are detected or suspected, the patient should be referred to a specialist in Fetal Medicine to get an additional anatomy scan and fetal echocardiography.	C
If the second-trimester anatomy scan is performed sufficiently and only iSUA is detected, a specialist fetal echocardiography is not necessary.	B
Invasive testing is <i>not</i> recommended on a routine basis in iSUA, because the risk of chromosomal anomalies is not increased.	B
Invasive testing with chromosomal evaluation (microarray) is recommended if associated malformations are detected (niSUA), because the risks of syndromes and chromosomal anomalies are substantially increased in this case.	B
It is recommended that fetuses with iSUA be followed with growth scans during the pregnancy, e.g., in GA week 28-32 and week 32-36.	B
If SGA/IUGR and/or other placental/cord anomalies are found, the controls should be more frequent.	B
For normal-weight fetuses with isolated SUA and normal insertion of the cord, no particular precautions during labor are needed and induction can be performed following the existing guidelines.	B
Children born with iSUA, where there are no other findings, do not need to be seen at the pediatric office.	B

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Abbreviations:

SUA	Single Umbilical Artery
iSUA	isolated Single Umbilical Artery
niSUA	non-isolated Single Umbilical Artery
SGA	Small for Gestational Age
FGR	Fetal Growth Restriction
IUGR	Intrauterine Growth Restriction
SGA	Small for Gestational Age
3VC	3 Vessel Cord
OR	Odds Ratio
UC	Umbilical Cord
FMF	The Fetal Medicine Foundation
DFMS	The Danish Fetal Medicine Society
DS	Downs Syndrome
OFT	Outflow Tracks
US	Ultrasound
GA	Gestational Age

Malformations:

Questions asked:

Investigate the frequency of SUA in populations, such as the Danes, who are evaluated with a first-trimester risk assessment for Down's Syndrome, and in populations who did not have a first-trimester risk assessment.

Investigate the frequency of associated malformations in fetuses having SUA in populations who have had a first-trimester risk assessment and in populations who did not.

Are further examinations required (fetal echo/US evaluation by a specialist in Fetal Medicine) in addition to the second-trimester anatomy scan in fetuses with isolated SUA?

Should the fetus be checked for SUA in the first trimester?

Summary of Evidence

Level of Evidence

The frequency of SUA is 0.5% in a low-risk background-population that had a first-trimester risk assessment. (12)	IV
iSUA constitutes 96% of SUA in a low-risk background population that had a first-trimester risk assessment. (12)	IV
The frequency of SUA in a population that did not have a first-trimester risk assessment depends on the population being investigated (i.e., high, low, or mixed risk for malformations) and when the investigation occurred (pre- or postnatally). SUA constitutes 0.44-1.3%. (8,14,22,38)	IIb-III
In a population with SUA, 4.3% is niSUA among those who did have a first-trimester risk assessment and 24.6% among those who did not. (11)	IIb
66 % of the heart malformations in niSUA were detected by the standard US-views of the heart described in FMF's recommendations. The heart malformations that were not detected by US were small and associated with good outcomes. (12)	IV
The observed accuracy of prenatal diagnosis of SUA in the second trimester is 99.92% (99.89-99.95%). (22)	IV

Clinical Recommendations

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It is recommended to check for SUA in the second trimester according to the DFMS guideline.	√
The second-trimester anatomy scan should be conducted by health care professionals who are educated and certified according to FMF standards. If SUA is detected, extra attention is warranted to detect possible associated malformations.	√
When SUA is detected, a thorough fetal heart scan is warranted. At a minimum, a 4-chamber view, OFT and 3-vessel view should be performed in sufficient detail.	C

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If the anatomy scan is not performed sufficiently (not all structures seen in detail), or if associated malformations (niSUA) are detected or suspected, the patient should be referred to a specialist in Fetal Medicine to get an additional anatomy scan and fetal echocardiography.	C
If the second-trimester anatomy scan is performed sufficiently and only iSUA is detected, a specialist fetal echocardiography is not necessary.	B
If niSUA is detected, the patient should be offered chromosomal testing (microarray).	B

Chromosomal Anomalies:

Questions asked:

Does SUA give an increased risk for chromosomal anomalies?

Is invasive testing warranted in isolated SUA detected in the second trimester?

Summary of Evidence

Level of Evidence

The risk of chromosomal anomalies is not increased in fetuses with iSUA; this applies for all pregnancies (both with and without a first-trimester risk assessment). (11)	IIa-IV
Fetuses/newborns with niSUA have 15 times increased risk of having a chromosomal anomaly compared to those who had a 3VC. (28)	IIb
niSUA is associated with trisomy 18, trisomy 13, triploidy, and Turners Syndrome, but not with trisomy 21 (11,17).	IIb

Clinical Recommendations

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Invasive testing is not recommended on a routine basis in iSUA, because the risk of chromosomal anomalies is not increased.	B
Invasive testing with chromosomal evaluation (microarray) is recommended if associated malformations are detected (niSUA), because the risk of syndromes and chromosomal anomalies are substantially increased in this case.	B

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SUA and Growth:

Questions asked:

Does isolated SUA increase the risk for intrauterine growth restriction?

Should the fetus be followed with growth scans during pregnancy, when iSUA is detected in the second trimester?

Summary of Evidence

Level of Evidence

Growth restriction in fetuses with iSUA is increased by at least a factor of 2 compared to a 3VC.	II-III
Placental/cord anomalies and amnion fluid anomalies are increased in fetuses with iSUA by a factor of 2-4.	II-III

Clinical Recommendations

Strength

It is recommended that fetuses with iSUA be followed with growth scans during the pregnancy, e.g., in GA week 28-32 and week 32-36.	B
If SGA/IUGR and/or other placental/cord anomalies are found, the controls should be more frequent.	B
For normal-weight fetuses with isolated SUA and normal insertion of the cord, no particular precautions during labor are needed and induction can be performed following the existing guidelines.	B

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Longterm outcome

Question asked:

How do those born with iSUA do in childhood?

Summary of Evidence

Level of Evidence

The long-term outcome for children with iSUA is the same as for kids born with 3 vessels in the umbilical cord. (9)	IIb
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Clinical Recommendations

Strength

Children born with iSUA, where there is no other findings, do not need to be seen at the pediatric office.	B
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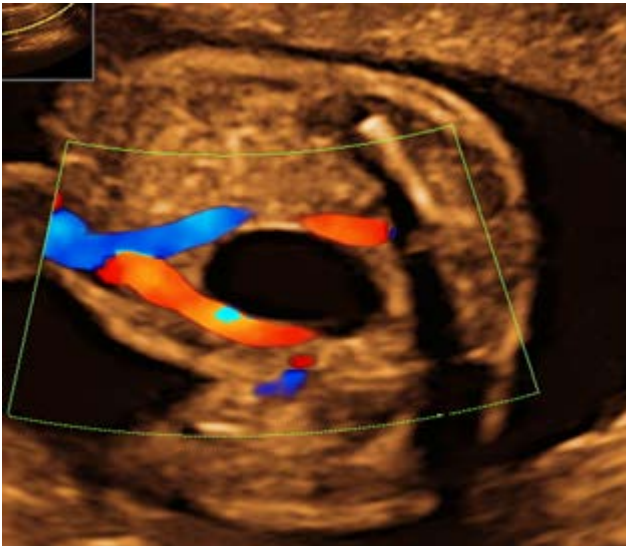
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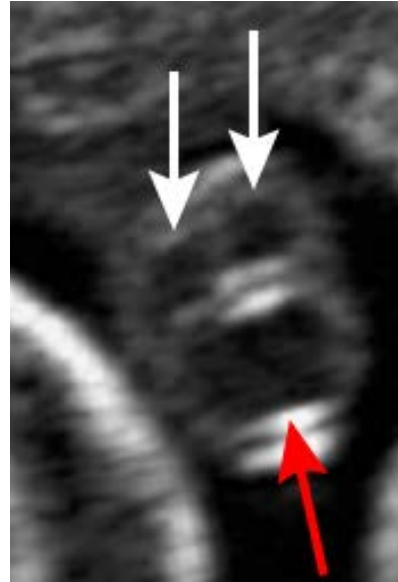
Appendix

Normal vessels and SUA at bladder level and in the umbilical cord (uc).

Normal:

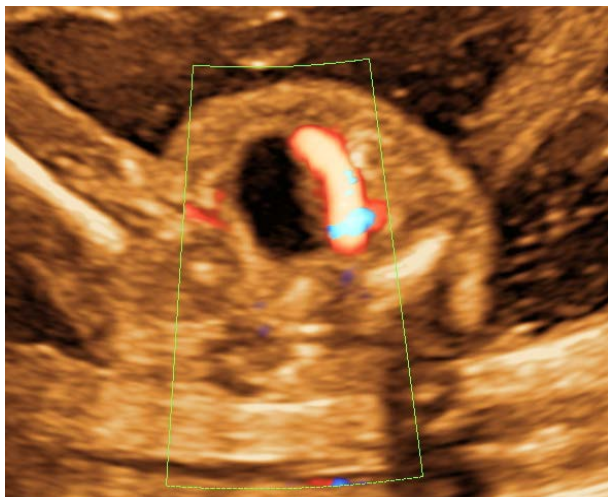


Picture 1: 2 aa. umbilicalis at the bladder level

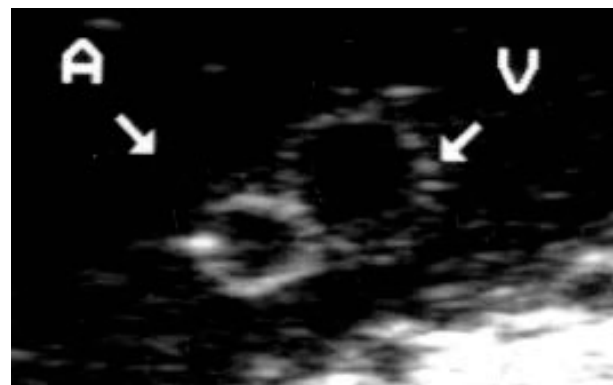


Picture 2: 2 arterier og 1 vene in uc

SUA:



Picture 3: Only 1 a. umbilicalis at bladder level



Picture 4: Only 1 artery (A) and 1 vene (V) in uc