POSTER SESSION

SMAkers®
Let's make the future of SMA together

Pregnancy with SMA
Our experience at the *Nemo Clinical Center*
Our pathway of care

Alice Zanolini, MD
Adult Neurologist - Centro Clinico Nemo Milano

ABSTRACT
There is only few evidence in literature of SMA woman with severe disability who became mothers; we have as physicians not a clear guideline to follow in these cases.
We report here our experience at the Nemo Center in Milan in the past years; we followed 3 SMA IIIa women with variable burden of disability during their first pregnancy experience.
Looking at our kind of care-taking and our experience's outcome we scheduled a reasonable pathway of care for pregnant women with SMA.

Background and Objectives
What we know from literature

- In SMA knowledge of the respiratory situation is essential for pregnancy planning.
- While successful pregnancies have been reported in women with vital capacities as low as 11%, there is an increased risk for miscarriages or stillbirths in patients with poor lung function which is still ill-defined.
- In SMA II and SMA III there is an increased preterm birth rate and most pregnancies end now via a planned caesarean section.
- However, there is still controversy about the appropriate options for anaesthesia and analgesia management in SMA patients, as information is limited.[1]

- The incidence of maternal and fetal complications is not higher in SMA women than in general population.
- Prematurity and operative delivery occurs more frequently in SMA pregnancies, for impaired muscular and ventilatory function.
- Regional anesthesia should be preferred; if general anesthesia is needed, depolarizing muscle blockers must be avoided.
- Respiratory function may decline during pregnancy and in this case non-invasive ventilation is the recommended treatment.
- Persistent exacerbation of weakness is reported in a significant proportion of women.[2]

Data
Our pregnant SMA women:

- 3 pts aged 35-38 when pregnant
- Type of SMA: IIIa with variable range of disability
- None of them requested medical assisted procreation
- We monitored them from 6 weeks of gestation until birth
- None of them was on nusinersen (#1: patient's choice due to invasive procedure; #2: spinal arthrodesis) nor risdiplam treatment (the latter was not available yet).

One of them was taking antidepressant drug (discontinued), the other two did not assume any chronic therapy besides vitamins.

Pts	Age at pregnancy (year)	*SMN2* copies	Loss of ambulation (yrs)	Column surgery (yrs)	FVC (%)	NIV	RULM pre/post	HMFSE pre/post	C-sec (weeks)	Anaesthesia
1	35 (2017)	2	17	No	46	For C-sec only	14/13	0/0	37	Epidural
2	38 (2018)	-	30	Yes (16)	57	For C-sec only	-	-	38	General
3	37 (2020)	3	11	Yes (11)	35	Yes (since 31 aa)	13/13	5/5	31	General (em.)

Pregnancy complications:
- Diabetes (#1, reversible)
- Anaemia (#2)
- Lumbar pain (#3, rev)

SMA complications:
- Worsening of motor function (#2)
- Worsening of respiratory function (#3, rev)
- Lumbar pain (#3, rev)

Outcomes:
- Hospitalization in NM Center (#2):
  - for nutritional support and blood transfusions (#1 – last 3 mos of pregnancy)
  - prepare for C-sec and help family with newborn management (#1)
- Caesarean section (#3 - #1 in emergency for mother hypotension)
  - #2 general anaesthesia for difficulties in epidural access and emergency procedure
- 3 healthy children, 2 boys and 1 girl
- None of the mothers did breastfeeding due to motor upper limb impairment and fatigue
- #1 patient had another baby 2 years after (2).

CONCLUSIONS
Our pathway of care for SMA mothers

Prior to pregnancy:
1. Neurological evaluation
➜ genetic counselling
➜ request multidisciplinary consultation
➜ leadership in pregnancy management
➜ treatment management (nusinersen, risdiplam)

2. Multidisciplinary approach
➜ gynecologist, pulmonologist, physiatrist, nutritionist, psychologist consultation

During pregnancy:
1. Monitoring schedule ➜ every 2 months together with gynecological follow-up
- respiratory evaluation
- motor/aids assessment

2. Interventions needed ➜ inpatient/outpatient fashion
- NIV adaptation/NIV parameters change
- Aids revision
- Management of complications (i.e. lumbar pain, anaemia, infections)
- Scheduling of caesarean section if necessary (recommended) – anaesthesiology team

Childbirth:
1. Anaesthesia:
- epidural vs sedation/general anaesthesia
➜ monitoring respiratory function/use of NIV – cough-assist

2. Provide a place in ICU

After childbirth:
1. Inpatient/outpatient management in a neuromuscular center
- management of pain and disability, posture
- educational for mother and family/caregivers
- psychological support (i.e. monitoring depression)

2. Long term multidisciplinary follow-up
➜ to monitor muscular and respiratory function stabilization/progression

A special THANK YOU
GRANDE OSPEDALE METROPOLITANO NIGUARDA di MILANO
Ostetricia e ginecologia - Dipartimento Materno Infantile
Anestesia e rianimazione 1 - Dipartimento Emergenza Urgenza – EAS

References
1. 179th ENMC international workshop: Pregnancy in women with neuromuscular disorders 5–7 November 2010, Naarden, The Netherlands. Fiona Norwood, Sabine Rudnik-Schöneborn. Published: September 22, 2011. DOI https://doi.org/10.1016/j.nmd.2011.05.009.
2. Pregnancy outcomes in women with spinal muscular atrophy: A review. Elena Abati, Stefania Corti. Published: March 02, 2018. DOI: https://doi.org/10.1016/j.jns.2018.03.001.

Cod. M-IT-00002289

Roche