Outcomes:

Primary outcomes: Provide accurate information to women with spina bifida about the impact of spina bifida on pregnancy and the impact of pregnancy on women with spina bifida

Secondary outcomes: Help women with spina bifida maximize sexual functioning

Tertiary outcomes: Understand management options during menopause in women with spina bifida, in relation to bone health and pelvic organ prolapse

Prenatal/infancy:

Clinical question:

1. How can we manage pelvic organ prolapse in toddlers with spina bifida?

Guidelines:

1. Pelvic organ prolapse can occur at any stage in life in women with spina bifida. In infants, the prolapse can be manually reduced (de Mola 1996)

Toddler:

Clinical question:

1. How can we manage pelvic organ prolapse in toddlers with spina bifida?

Guidelines

1. Pelvic organ prolapse can occur at any stage in life in women with spina bifida. In toddlers, the prolapse can be manually reduced (de Mola 1996)

Pre School

Clinical question:

1. How can we manage pelvic organ prolapse in preschool children with spina bifida?

Guidelines

1. Pelvic organ prolapse can occur at any stage in life in women with spina bifida. In preschool children, the prolapse can be manually reduced (de Mola 1996)

School Age:

Clinical question:

1. How can we manage pelvic organ prolapse in school-age children with spina bifida?
Guidelines

1. Pelvic organ prolapse can occur at any stage in life in women with spina bifida. In school-aged children, the prolapse can be manually reduced (de Mola 1996)

Clinical question:

1. What are the specific concerns regarding osteoporosis for school age children with SB?

Guidelines:

1. Children with osteoporosis should maintain appropriate vitamin D and calcium levels. (Boyce 2014)
2. Children with osteoporosis should engage in physical activity, especially weight bearing exercise. (Boyce 2014)
3. Children with severely compromised bone health should avoid activities associated with a high risk of fracture (Boyce 2014)

Teenage:

Clinical question:

1. How can we manage pelvic organ prolapse in adolescents with spina bifida?

Guidelines:

1. Pelvic organ prolapse can occur at any stage in life in women with spina bifida. In teens, the prolapse can be manually reduced or a pessary can be used. Surgical options are also available and should be discussed in terms of fertility preservation with a urogynecologist (de Mola 1996)

Clinical question:

1. What are the specific concerns regarding osteoporosis for adolescents with SB?

Guidelines:

1. Adolescents with osteoporosis should maintain appropriate vitamin D and Calcium levels. (Boyce 2014)
2. Adolescents with osteoporosis should engage in physical activity, especially weight bearing exercise. (Boyce 2014)
3. Adolescents with severely compromised bone health should avoid activities associated with a high risk of fracture (Boyce 2014)

Adult:

Clinical question:
1. How can we maximize the physical sexual functioning of women with spina bifida, including orgasm, lubrication and engorgement?

Guidelines:
1. There is no specific literature on this topic and requires further research

Clinical question:
1. What are the impacts of pregnancy on the global physiology of the SB patient?

Guidelines:

**Breathing**

1. Dyspnea can occur pregnancy when there is an associated kyphoscoliosis deformity. In women with kyphosis/scoliosis, pulmonary function testing should be done at least once during pregnancy. All women with kyphosis, scoliosis or dysmobility should be asked about and shortness of breath should be asked at each antenatal visit, and pulmonary function testing or assessment for pulmonary embolism undertaken as indicated (Blasi 2012)

**Bowels**

1. Bowel care should be discussed early in pregnancy, as pregnancy can worsen constipation. A diet high in fiber and use of stool softeners are safe in pregnancy.
2. Consultation with a dietician should be considered to maximize dietary methods of alleviating constipation.
3. Suspected bowel obstruction should be managed by an interdisciplinary team consisting of a general surgeon and obstetrician.

**Shunt**

1. The enlarging uterus can cause a shunt malfunction by increasing the intraabdominal pressure. Signs of increased pressure, headache, nausea, vomiting should be reviewed at each prenatal visit. Wang 2013
2. Signs of shunt malfunction should be managed with an interdisciplinary team consisting of neurosurgeon, obstetrician and anesthetist. Wang 2013
3. Shunt malfunction can appear similar to preeclampsia. A thorough work up for both preeclampsia and shunt obstruction should be done if a pregnant woman with a shunt has nausea, vomiting, headache or neurological symptoms Wang 2013

**Deep Vein Thrombosis Risk**

1. Women who have decreased mobility have an increased risk of deep vein thrombosis and pulmonary embolism in pregnancy. Thromboprophylaxis should be prescribed for women with limited mobility and those who use wheelchairs.
**Bladder and renal function**

1. Regular urinalysis and urine culture should be performed throughout pregnancy and infections promptly treated as urinary tract infections are common during pregnancy in mothers with spina bifida.
2. Baseline renal assessment should be made early in pregnancy.
3. Women who already have evidence of renal disease have a risk of decreased renal function in pregnancy and should be managed in conjunction with a nephrologist.
4. Women who have renal disease have an increased risk of preeclampsia and intrauterine growth restriction, and should have intensified maternal and fetal monitoring.
5. Stomal appliances can develop poor conduit drainage as the uterus grows. Women should be asked about the function of appliances at each visit. Poor conduit drainage may require stomal catheterization.
6. Patients with continent urinary diversions may develop increased incontinence or difficulties in intermittent self-catheterization, which may require temporary insertion of an indwelling catheter.

Clinical Question:

1. What are considerations for types of delivery for the SB patient?

Guidelines:

1. Vaginal delivery is possible, however severe spinal and pelvic skeletal deformities may prevent vaginal birth.
2. Vaginal deliveries are recommended in women with VP shunts with a shortened second stage to decrease elevation of intracranial pressure. (Hwanf 2010)
3. Some women may be as aware of labour contractions, and therefore should be taught to palpate for hardening of the belly, and observe for rupture of membranes, and signs of autonomic dysreflexia.
4. Autonomic dysreflexia can be triggered by labour in women with a lesion above T6. This condition can be life threatening and women experiencing any signs or symptoms should present to hospital immediately. (Dakhil-Jerew 2008)
5. Autonomic dysreflexia has a clinical overlap with preeclampsia, and pregnant women with a lesion above T6 suspicious for autonomic dysreflexia or preeclampsia should be evaluated for both.
6. Cesarean section in patients with previous lower urinary tract surgery are not contraindicated, however may be hazardous. Intestinal and omental adhesions to the lower uterine segment may necessitate classic upper segment section. The decision between a planned caesarian section with available urology back up vs a plan for trial of vaginal delivery with the associated risks of emergency caesarian sections should be made with a multidisciplinary team consisting of an anesthetist, urologist and obstetrician, taking into account patient goals and preferences.
7. Cesarean section is recommended for women with vesical neck reconstruction or artificial sphincter placement to protect continence (Thomas 2009)
8. Women with bladder extrophy and spina bifida should undergo a planned cesarean section to preserve continence and decrease risk of pelvic organ prolapse. (Thomas 2009)

9. Pregnancy in of itself can exacerbate existing pelvic organ prolapse. Vaginal delivery will likely cause worsening of pelvic organ prolapse. Plan for mode of delivery should take into account the impact of this worsening and the possible need for subsequent surgery in consultation with an obstetrician and urogynecologist, taking into account patient preferences(Thomas 2009)

10. Spina bifida can be associated with congenital renal malformations, such as horseshoe kidney and pelvic kidney. The surgeon needs to be aware of renal anatomy prior to C-section and may require consultation with a urologist if C-section is required.


12. Women with a cesarean delivery and shunt should be observed following delivery for shunt related neurological changes as these can happen in the immediate post-partum time frame, and can be associated with blood clots obstructing the shunt. (Hwang 2010)

13. If a C-section must be undertaken for obstetric reasons in a woman with a VP shunt, careful attention to hemostasis and irrigation should be used to decrease the risk of fibrin depositing in the shunt (Hwang 2010)

Clinical Question

1. What is the impact of SB on pregnancy?

Guidelines:

1. Spina bifida is not a contraindication to epidural anesthesia; however each woman should have an anesthesia consultation prior to delivery to discuss risks and benefits of regional vs general anesthesia. (Tidmarsh 1998)

2. It appears that there is an increased risk for preterm birth in women with spina bifida. This may be related to the increased risk of mullarian abnormalities, or an intrinsic increased risk of preterm birth seen in women with spinal cord injuries. Signs and symptoms of preterm birth should be discussed with each woman in the context of her sensory abilities.

3. Transvaginal ultrasound for cervical length assessment q 2-4 weeks should be considered to assess risk of preterm birth. In women with a history of autonomic dysreflexia, this should be done transabdominally.

4. Cervical shortening at a previable gestational age can be managed with cerclage (Brown 2013) or progesterone as indicated (Farine et al 2008)

5. Administration of betamethasone for fetal lung maturity should be provided if there are signs or symptoms of preterm birth after age of viability. (Crane 2003)
Clinical question:

1. Is breastfeeding impacted by SB and if so how?

Guidelines

2. There is no literature specifically on breast feeding in the context of spina bifida.
3. Women who wish to breast feed should be encouraged to do so and be provided with support from a lactation consultant.

Clinical question:

1. How can we manage pelvic organ prolapse in adults with spina bifida?

Guidelines:

1. Pelvic organ prolapse can occur at any stage in life in women with spina bifida. The use of pessaries and surgical repair should be considered in consultation with a urogynecologist. (de Mola 1996)

Clinical Question:

1. What are the specific concerns regarding osteoporosis for women with SB?

Guidelines:

1. Bisphosphonates may increase bone mineral density and should be considered in adult women with spina bifida (Smith 2011)
2. Vitamin D supplementation may reduce the rate of bone mineral density decline (Smith 2011)

Research Gaps

1. There is no literature specifically on breast feeding in the context of spina bifida.
2. How can we maximize the physical sexual functioning of women with spina bifida, including orgasm, lubrication and engorgement?

References


Hatch TR, Steinberg RW, Davis LE. Successful term delivery by caesarean section in a patient with a continent ileocaecal urinary reservoir. J Urol 1991;146:11–1112
