Oppfølging av lipomer i spinalkanalen

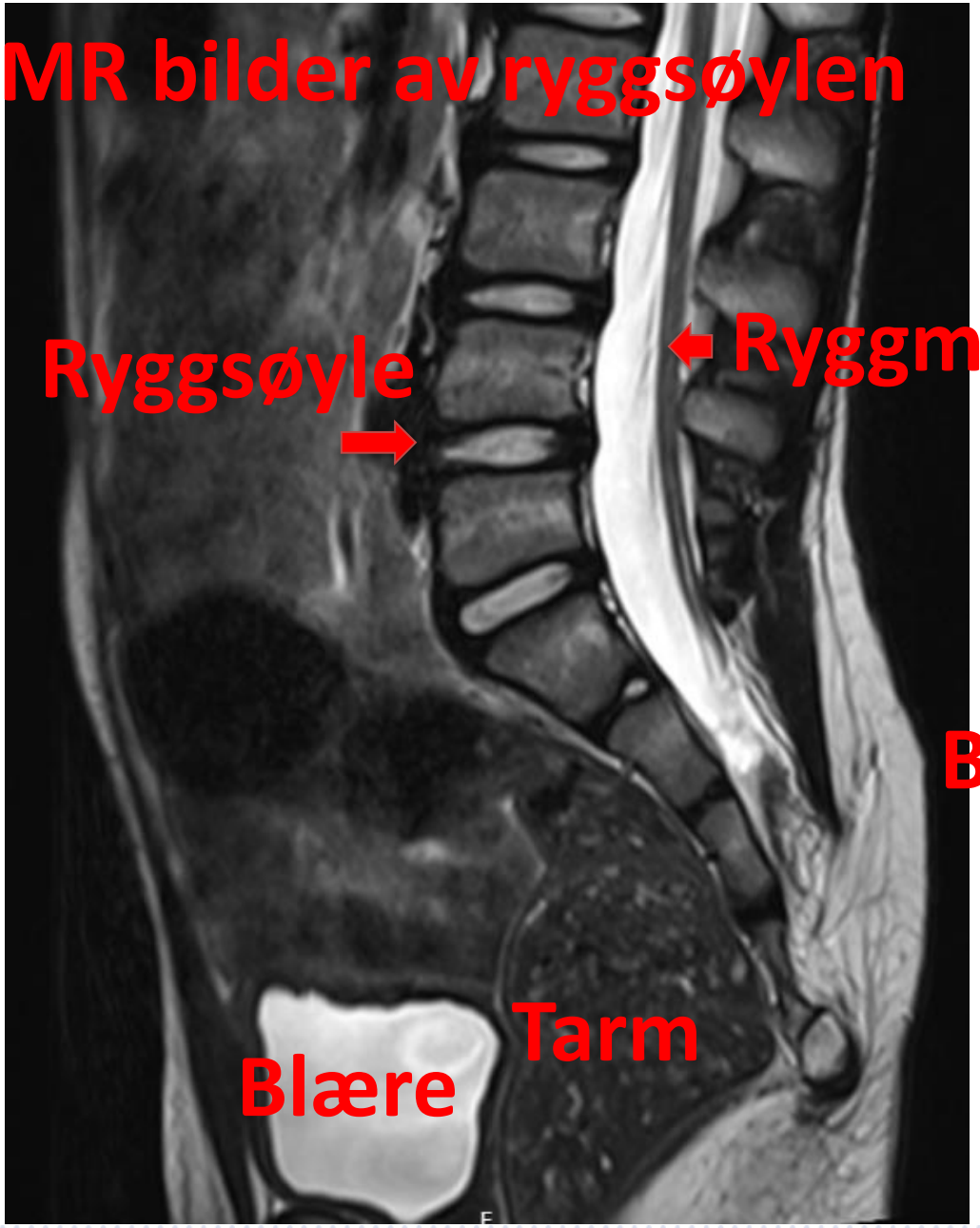
Petra Aden

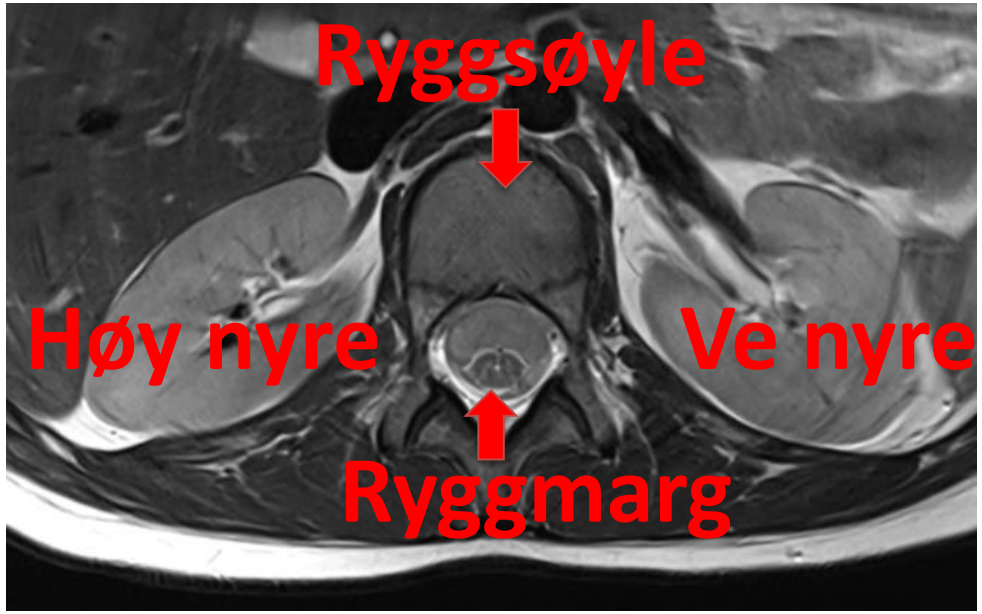
Nasjonalt senter for sjeldne diagnoser – TRS

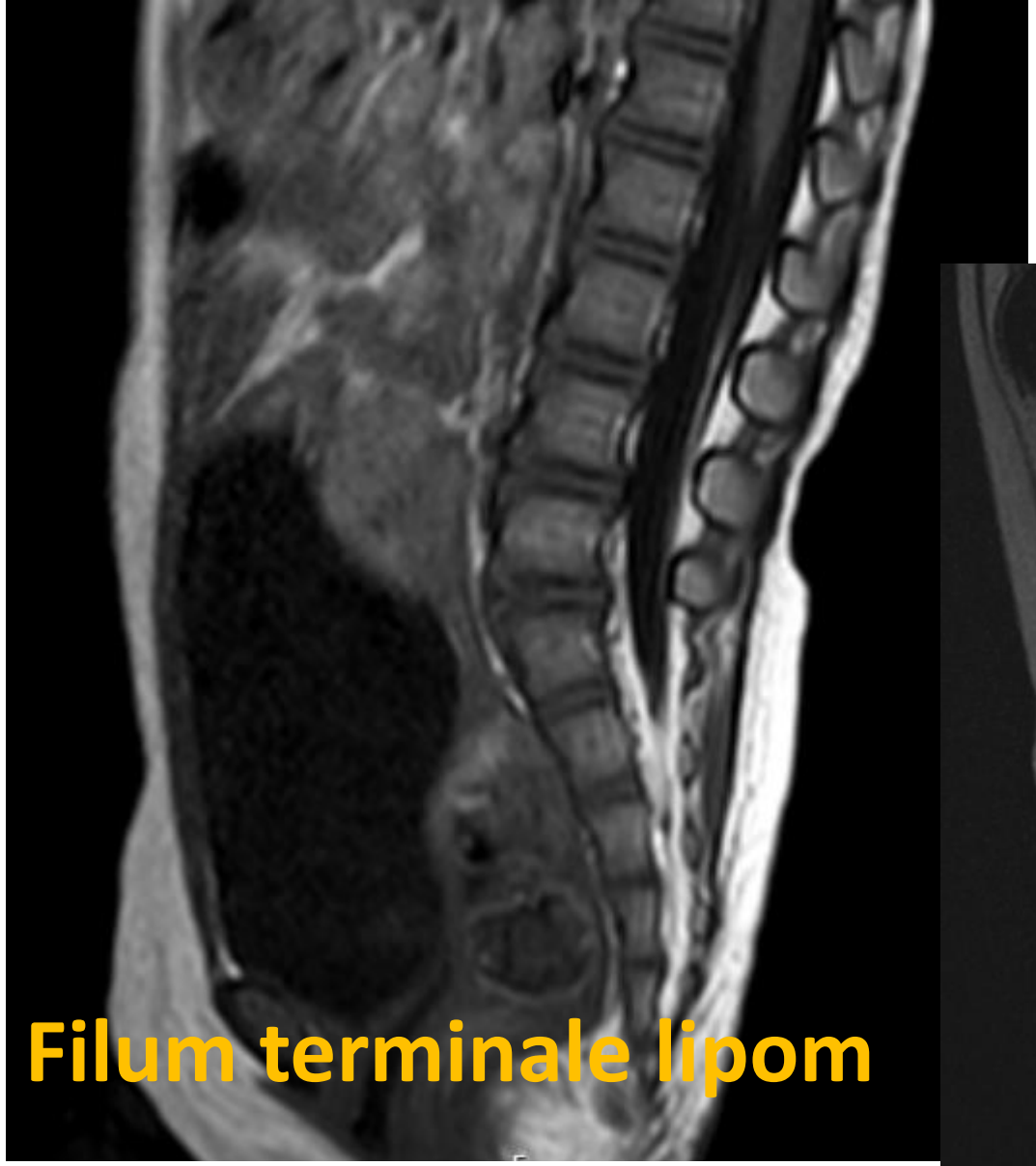
Oslo, Norge

- Neural tube defects
  - Neural tube closure defect
    - Anomaly of the filum terminale
    - Spinal cord lipoma (SCL)
      - Intramedullary non-dysraphic SCL
      - Dysraphic SCL
        - Dorsal SCL
        - Conus SCL
          - Chaotic SCL
          - extramedullary SCL
    - Spina bifida and other spinal dysraphisms
      - Closed spinal dysraphism
        - Myelocystocele
        - Dermal sinus
        - Limited dorsal myeloschisis
      - Open spinal dysraphism (OSD)
        - Open spinal dysraphism with myelomeningocele
        - Myeloschisis
  - Malformation of the neurenteric canal, spinal cord and column
    - Split cord malformation (SCM)
    - Caudal regression syndrome
    - Primary syringomyelia
    - Spinal segmental dysgenesis

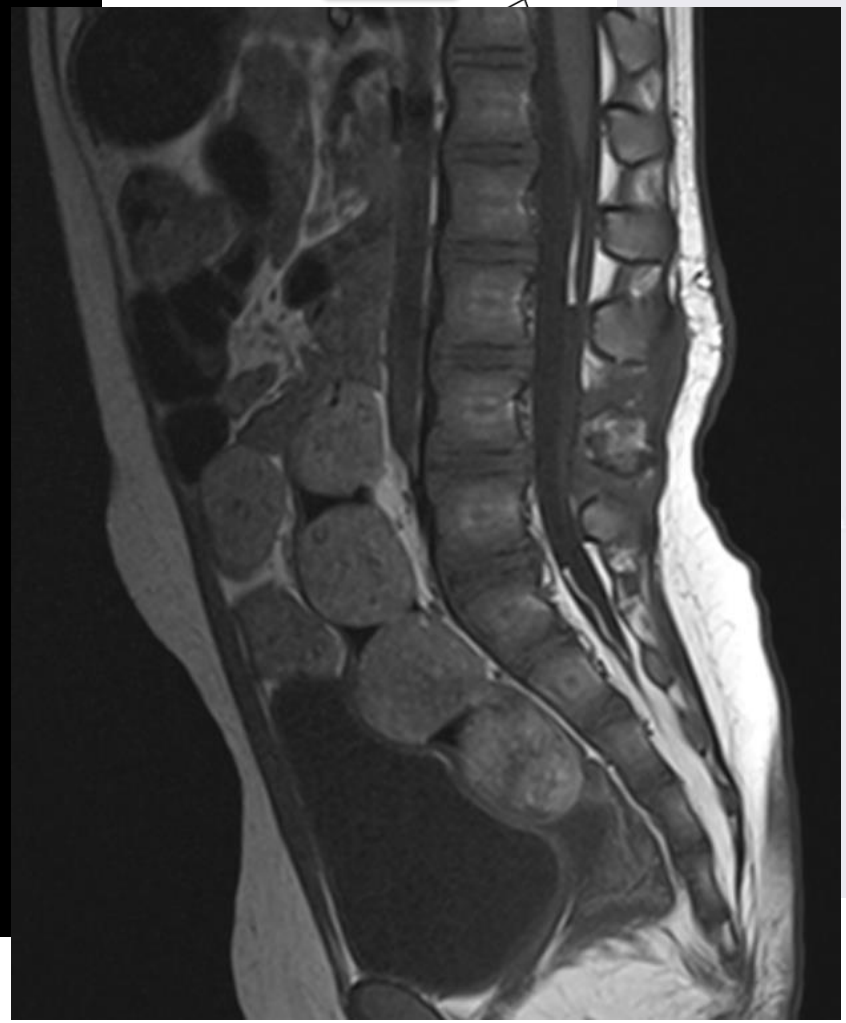
MR bilder av ryggsøylen

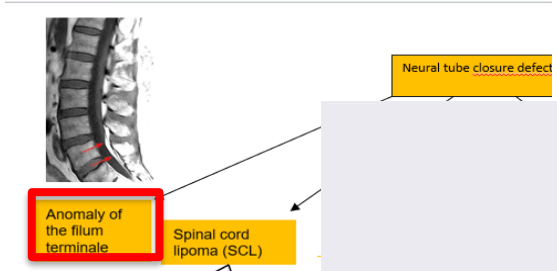


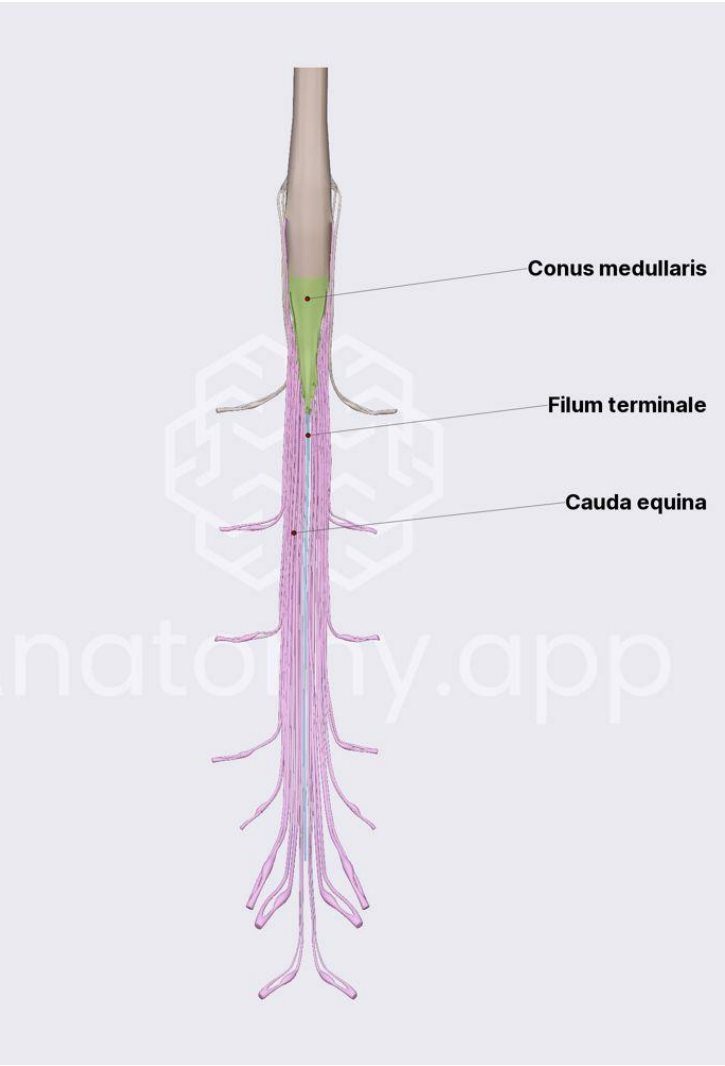




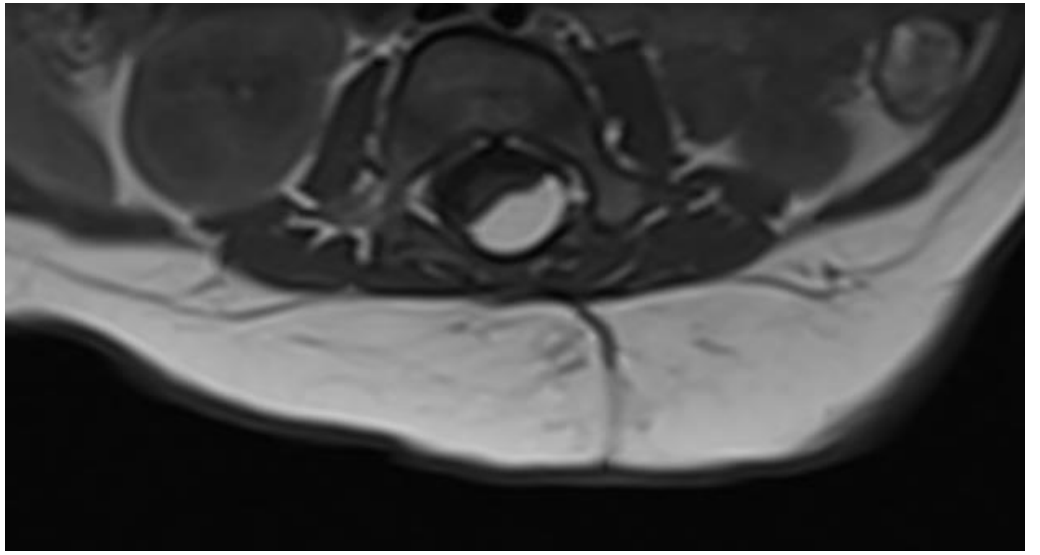
Filum terminale lipom

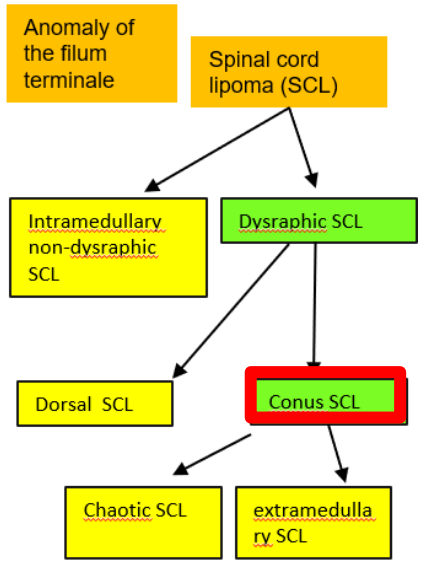






Conus Lipoma



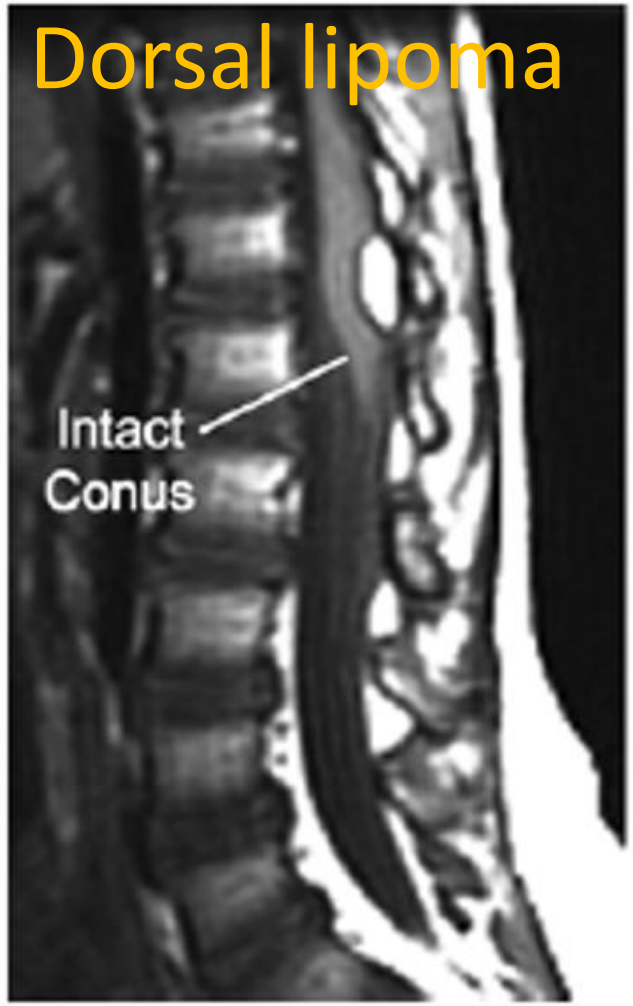


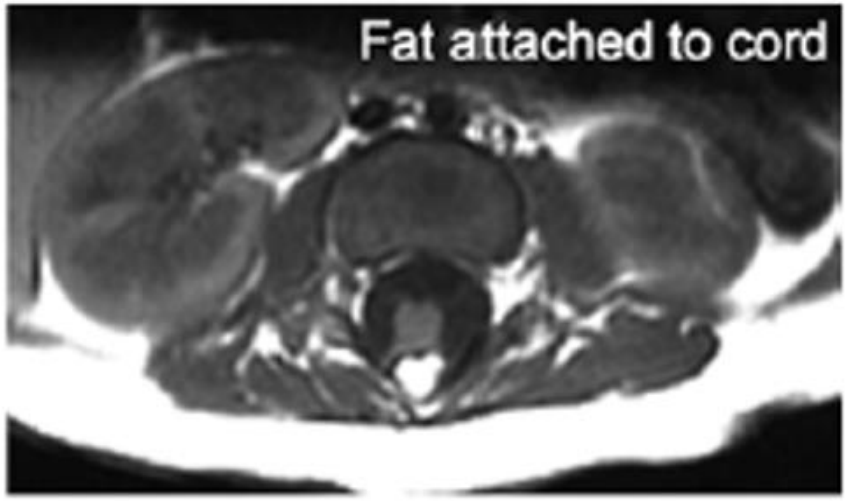
Suggest an update

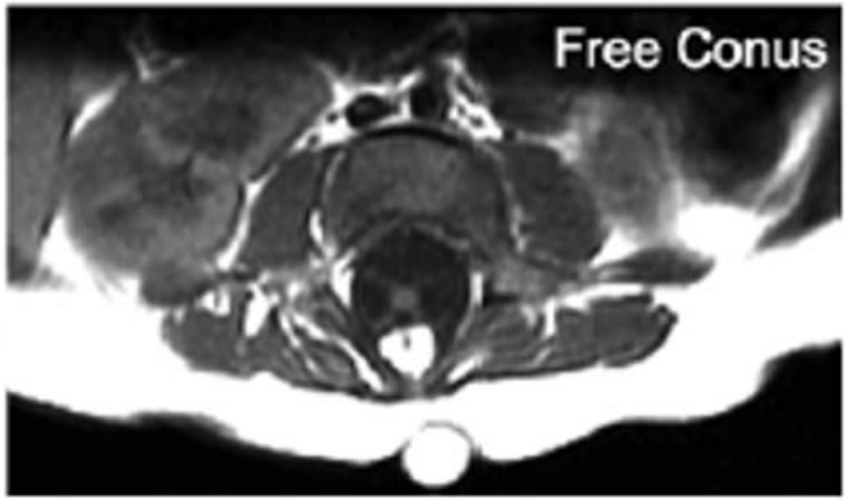
Disease definition

A rare lipomatous, dysraphic malformation characterized by lipoma located wholly or partially at the conus.

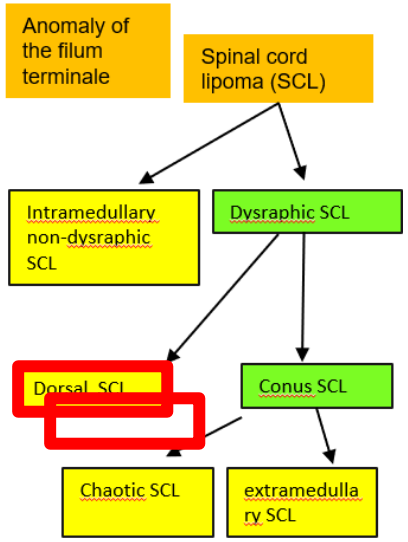
Dorsal lipoma







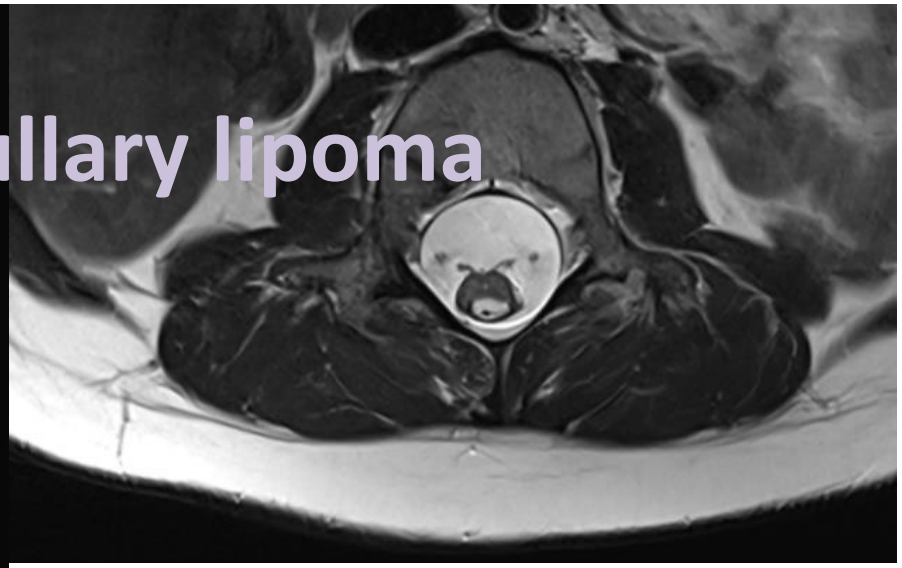
Suggest an update

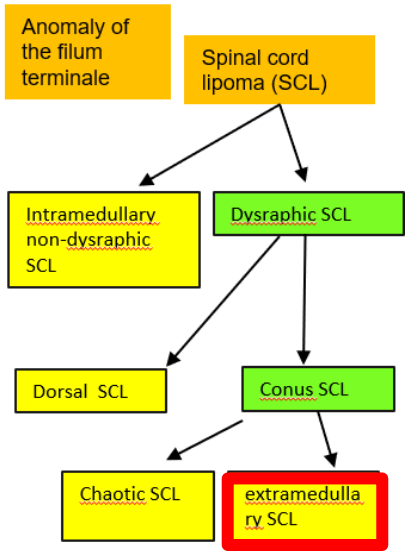


Disease definition

A rare lipomatous, dysraphic malformation characterized by attachment to the dorsal surface of the spinal cord but not extending to the conus. It can be associated with others features such as a stalk and vertebral bone abnormalities.

Transitional extramedullary lipoma





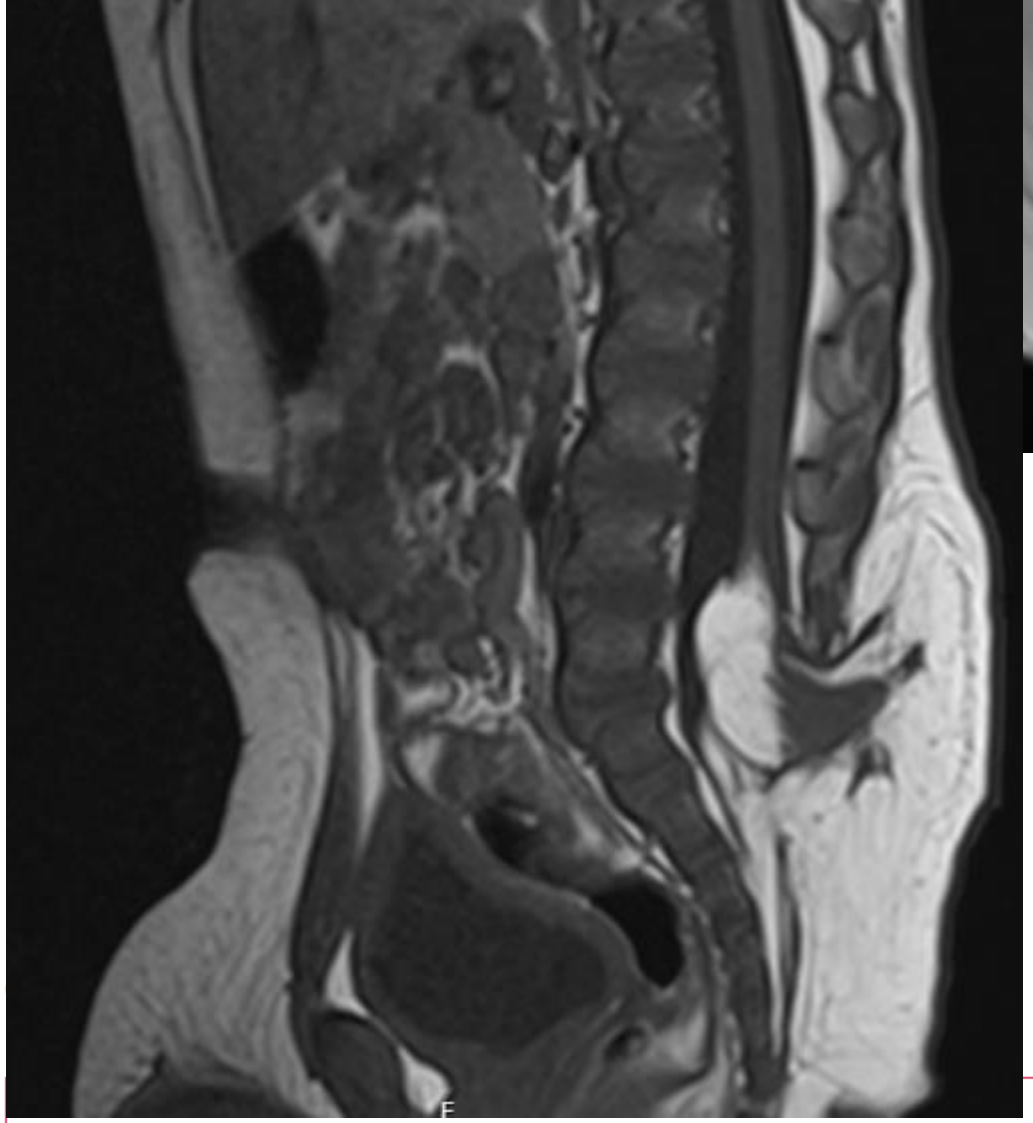
Transitional extramedullary conus spinal cord lipoma

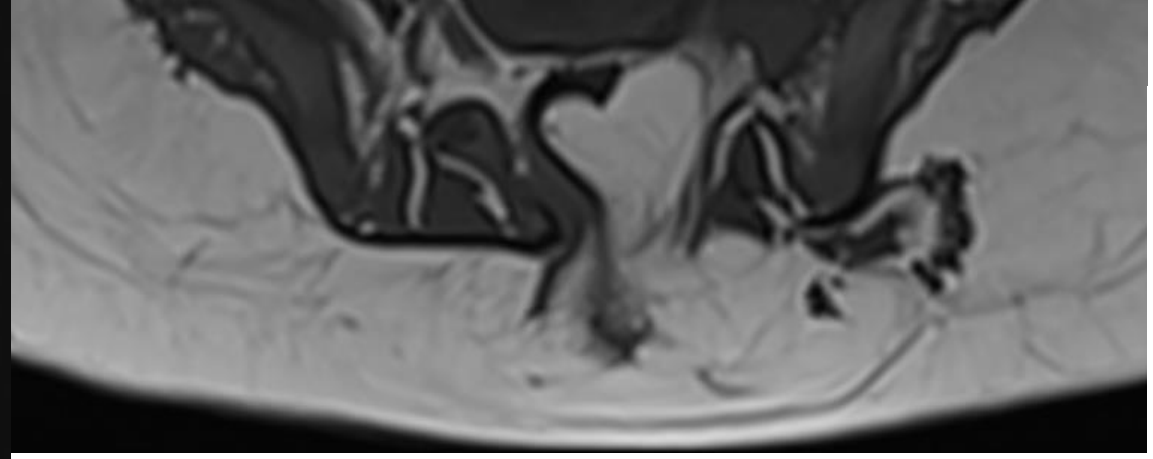
Suggest an update

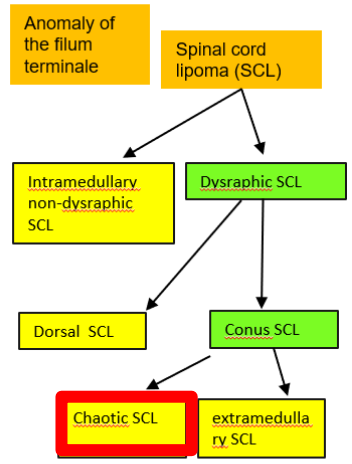
Disease definition

A rare form of extramedullary conus region spinal cord lipoma characterized by a location at the posterior surface of the conus and its tip. i.e-"transitional" between posterior and terminal conus region spinal cord lipoma.

NASJONALT SENTER FOR SJELDNE DIAGNOSER







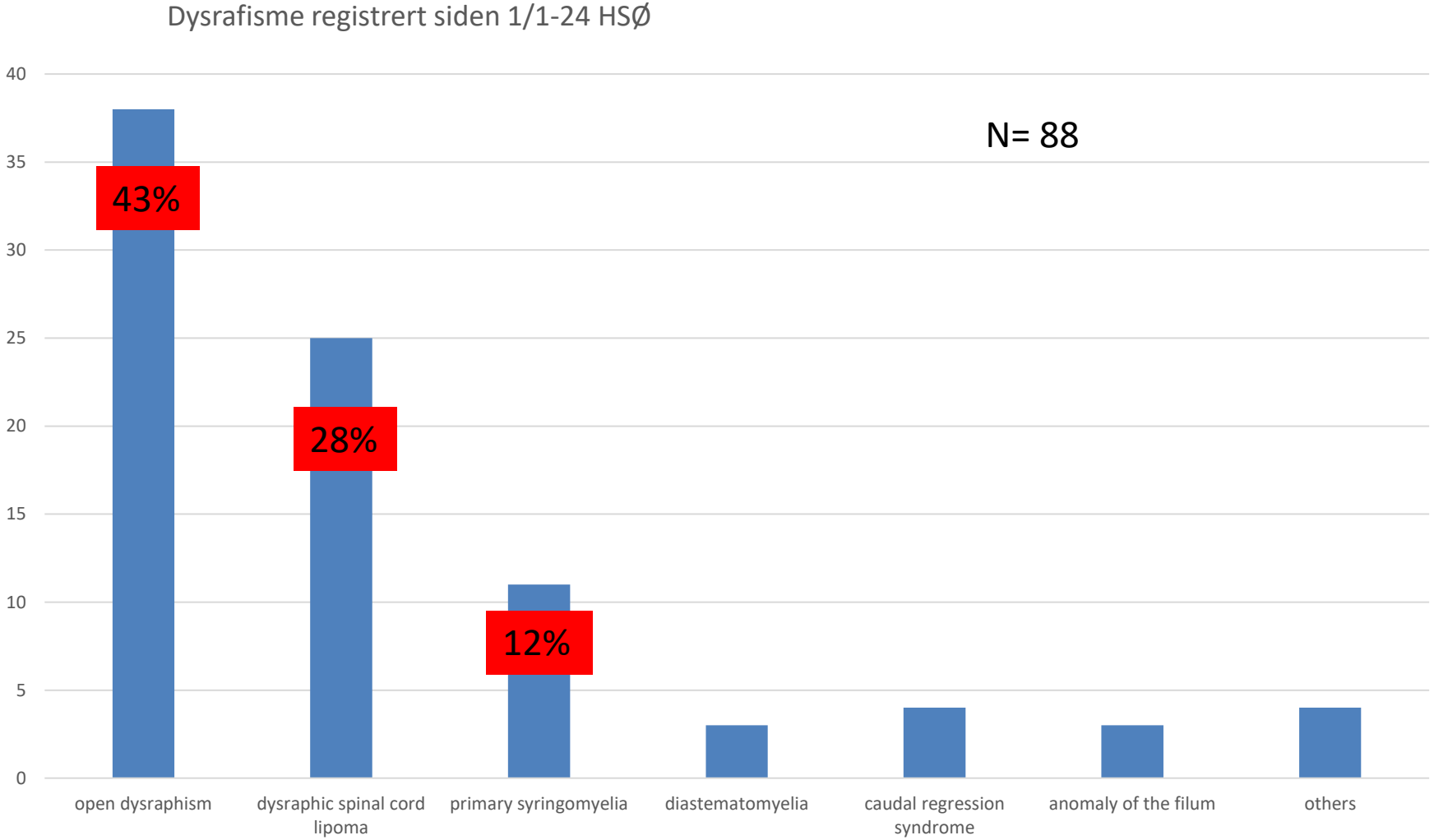
Suggest an update

Disease definition

A rare dysraphic spinal cord lipoma characterized by the lipomatous mass extending ventrally to the dorsal root entry zone, indicating a more severe malformation of the spinal cord. The diagnosis can be suggested on imaging but usually confirmed during surgery.

Epidemiologi

0,3-0,6 per 10.000 levendfødte *(Sarris 2012)*



Risikofaktorer hos mor

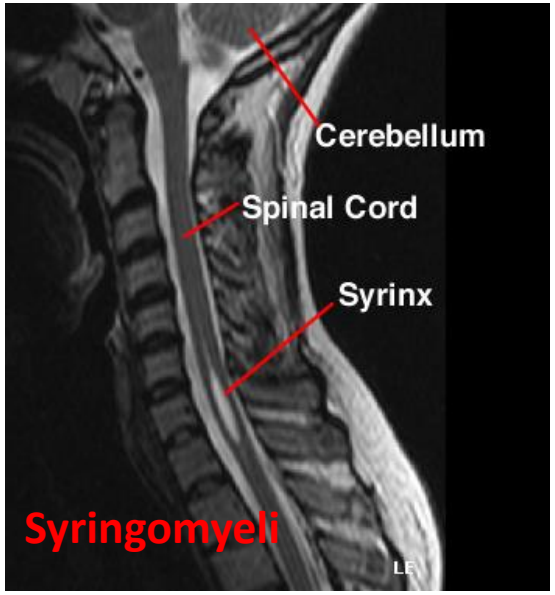
- Vitamin B12-mangel
- Diabetes hos mor
- Fedme hos mor
- Bruk av antiepileptika under graviditet
- Folat?

Problemstillinger ved lipomer

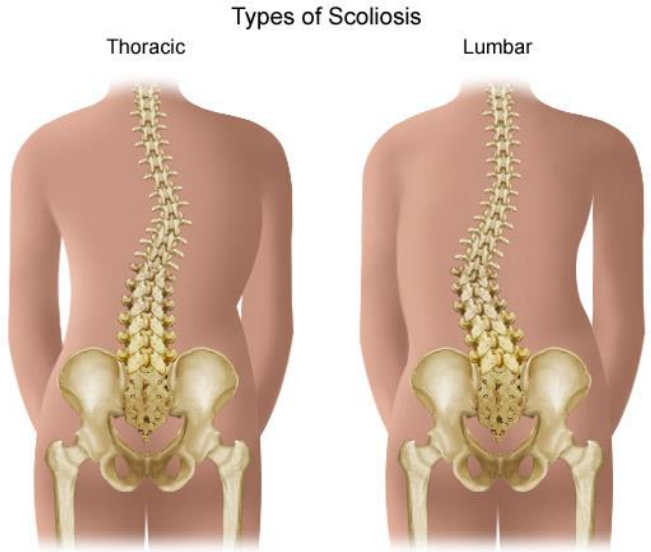
- Syringomyeli
- Tjoret marg
- Smerter I ryggen
- Motoriske problemer
- Blære/tarmproblemer

Problemstillinger ved lipomer - oversikt

- Syringomyeli
- Tjoret marg
- Smerter I ryggen
- Skoliose
- Motoriske problemer
- Blære/tarmproblemer



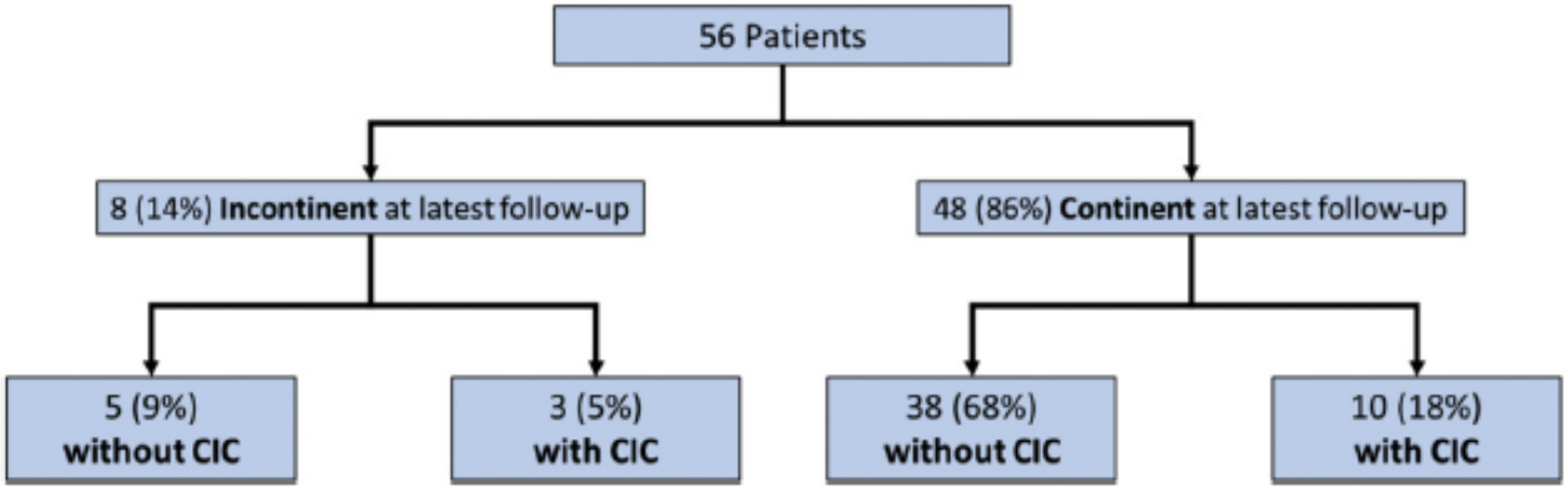
Tjoret marg



Journal of Pediatric Urology (2017) 13, 371.e1–371.e8

Lipomyelomeningocele for the urologist: Should we view it the same as myelomeningocele?

E.B. Yerkes, C. Halline, G. Yoshiba, T.A. Meyer, I. Rosoklija,
R. Bowman, D. McLone, E.Y. Cheng



Summary Figure Continence and CIC outcomes.

Risikofaktorer for inkontinens

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	Latest follow-up continence status		*P*-value
	Continent	Incontinent	
Surgery			1.0
Complete	36 (86%)	6 (14%)	
Partial	12 (86%)	2 (14%)	
Lipoma type			0.75
Dorsal	21 (84%)	4 (16%)	
Distal	14 (93%)	1 (7%)	
Transitional	12 (80%)	3 (20%)	
Chaotic	1 (100%)	0	
Pre-operative urodynamics ($n = 28$)			0.99
Low risk	5 (100%)	0	
Intermediate	18 (86%)	3 (14%)	
High risk	2 (100%)	0	

Risikofaktorer for RIK

Journal of Pediatric Urology (2017) 13, 371.e1–371.e8

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Table 3 Latest follow-up CIC status.

	Latest follow-up CIC status		*P*-value
	Not on CIC	Yes on CIC	
Surgery			0.001
Complete	37 (88%)	5 (12%)	
Partial	6 (43%)	8 (57%)	
Lipoma type			0.027
Dorsal	23 (92%)	2 (8%)	
Distal	11 (73%)	4 (27%)	
Transitional	8 (53%)	7 (47%)	
Chaotic	1 (100%)	0	
Pre-operative urodynamics ($n = 28$)			0.032
Low risk	5 (100%)	0	
Intermediate	18 (86%)	3 (14%)	
High risk	0	2 (100%)	

- Anticholinergics 39,2%
- Beta 3 agonist 10,8%
- Alpha blockers 10,8%

- DMSA 10,9% kidney damage

Neurourology Urodynamics

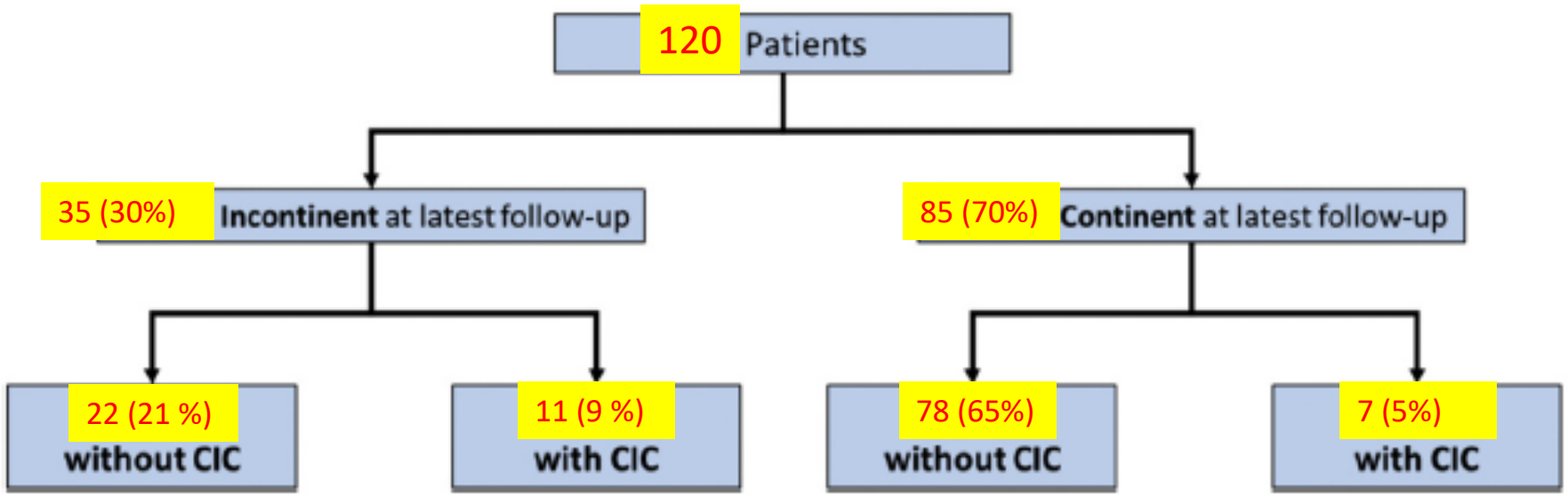
CLINICAL ARTICLE | Full Access

Analysis of factors affecting permanent clean intermittent catheterization and bladder function after primary neurosurgical repair of lipomyelomeningocele

Correction(s) for this article

Corrigendum
Volume 42, Issue 4, Neurourology and Urodynamics | pages: 888-888 |
First Published online: February 23, 2023

Yoonhye Ji RN, KAPN, Sang Woon Kim MD, Sang Won Han MD, Yong Seung Lee MD



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Risikofaktorer for inkontinens

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		Continence group ($n = 85$)	Incontinence group ($n = 35$)	p Value
Sex, n (%)	Female:male	44 (51.8%):41 (48.2%)	19 (54.3%):16 (45.7%)	0.802
Type of LMMC, n (%)	Caudal	40 (47.1%)	11 (31.4%)	0.047
	Dorsal	34 (40.0%)	13 (37.1%)	
	Transitional or chaotic	11 (12.9%)	11 (31.4%)	
Preoperative UD score	T0 UD score, mean (SD)	2.90 (2.369)	3.71 (2.244)	0.084
	≥5 (cutoff), n (%)	17 (20.2%)	10 (28.6%)	0.323

Patient characteristics	Incontinence Model 3 OR (95% CI)	p Value
Type of LMMC		
Caudal	ref[1]	
Dorsal	1.36 (0.54–3.49)	0.525
Transitional or chaotic	3.45 (1.15–10.38)	0.028
Age 36 months or older	0.86 (0.21–3.45)	0.827
T0 UD score ≥5	1.37 (0.50–3.76)	0.540

Risikofaktorer for RIK

		Non-CIC group ($n = 98$)	CIC group ($n = 22$)	p Value
Sex, n (%)	Female:male	52 (53.1%):46 (46.9%)	11 (50.0%):11 (50.5%)	0.795
Type of LMMC, n (%)	Caudal	46 (46.9%)	5 (22.7%)	<0.001
	Dorsal	43 (43.9%)	4 (18.2%)	
	Transitional or chaotic	9 (9.2%)	13 (59.1%)	
Preoperative UD score	T0 UD score, mean (SD)	2.76 (2.08)	4.82 (2.77)	0.003
	≥5 (cutoff), n (%)	16 (16.5%)	11 (50.0%)	0.002

Patient characteristics	CIC Model 1 OR (95% CI)	p Value
Type of LMMC		
Caudal	ref[1]	
Dorsal	1.69 (0.34–8.43)	0.522
Transitional or chaotic	13.82 (3.18–60.14)	<0.001
Age 36 months or older	7.76 (1.44–41.69)	0.017
T0 UD score ≥5	2.68 (0.75–9.54)	0.128

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Oppsummering

- 15-30 % Inkontinens for urin
- 15-25 % RIK

- Type av spinal lipom
- Alder for løsning av tjoret marg/lipom
- Type av løsning av tjoret marg/lipom
- Preoperativ høy-risk blære

Oppfølging for all spinal dysrafisme

- Henvisning til habilitering
- Årlig nevrologisk undersøkelse
- Årlig UL urinveier
- Årlig uro/tarmanamnese
  - Forstoppelse
  - Miksjonsliste (miksjonmengde/frekvens/kontinens)
  - Miksjonstrang
  - UVI?
- Eventuell henvisning til spesialundersøkelser (miksjonsobservasjon, MCUG, cystometri)

Avvik ved oppfølging av lipom

Undersøkelser	Nyfødt	3–4 md.	1 år	Årlig	Kommentar
Cystometri		x	x	x	Kun hvert 2.år v/lavtrykksblære eller uten infeksjoner
Miksjonsobs m/resturinbest	x	x	x	x	Resturinbest v/UL eller kateterisering
Urografi		(x)			Ved mistanke om misdannelser i urinveier
UL-urinveier evt. med resturinbestemmelse	x		x	x	Nyrestørrelse og –tilvekst. Nyrebekken-, ureterpatologi, hydronefrose
MUCG	x				Gjentas etter indikasjon som påvist refluks, pyelonefritt.
DMSA-scintigrafi			x	(x)	Ved indikasjon som hyppige pyelonefritter eller operative tiltak
MAG-3 renografi clearance					Ved mistanke om forskjell i funksjon mellom nyrene og avløpshinder
Bleieveietest		x	x	x	
Cystatin C				x	
GFR				(x)	Ved tegn på nyresvikt eller komplisert nevrogen blære, ellers ved 10 og 17 års alder.
U-stix og -dyrkning	x	x	x		Månedlig første leveår, så v/behov

Ikke første valg ved lipom

Helst ved etablert RIK

Bare ved indikasjon for cystometri eller hyppig UVI

Gjennomføring ved behov for alle type dysrafismer

Varselstegn

- Økende resturin (miksjonsobservasjon, flowmetri)
- Nyoppstått urinveisinfeksjoner
- Trabekulær blære (UL urinveier, MR columna)
- Høy trykk i blæren (cystometri)
- Nyoppstått vesikouretral refluks (UL urinveier, MCUG)
- Nyoppstått obstipasjon
- Ny tilkomne motoriske/nevrologiske symptomer
- Ofte en prosess over år
- Jo eldre barnet jo mer sannsynlig
- Jo senere løsning jo mer sannsynlig

Klinisk undersøkelse

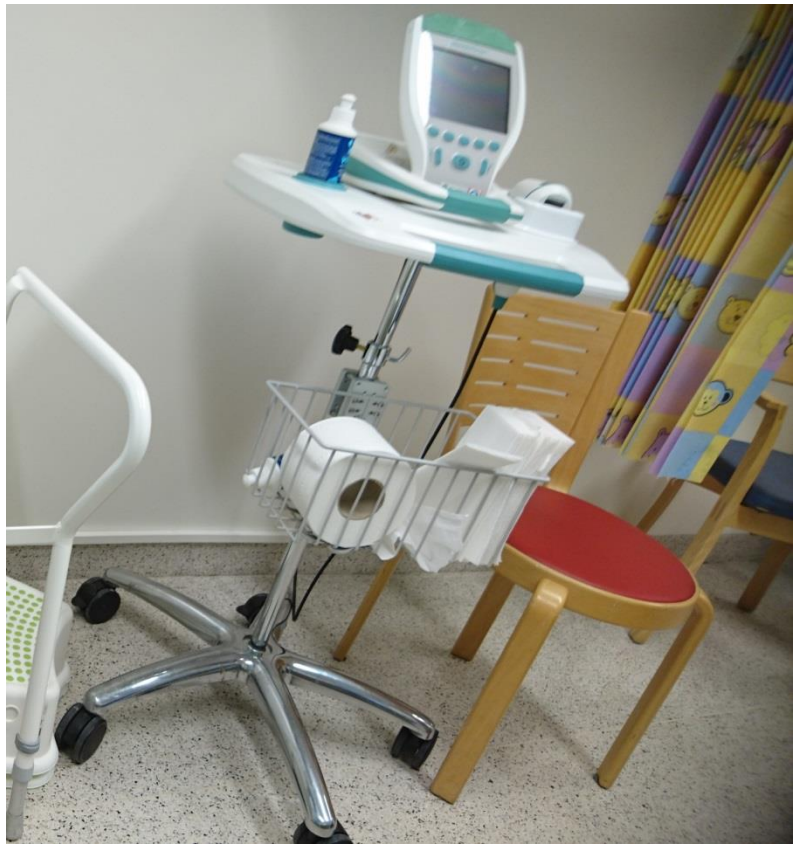
- Nevrologisk undersøkelse
  - Reflekser
  - Tonus
  - Catch
  - Feilstillinger
  - Sensibilitet
  - Gang/løpemønster
  - Skoliose

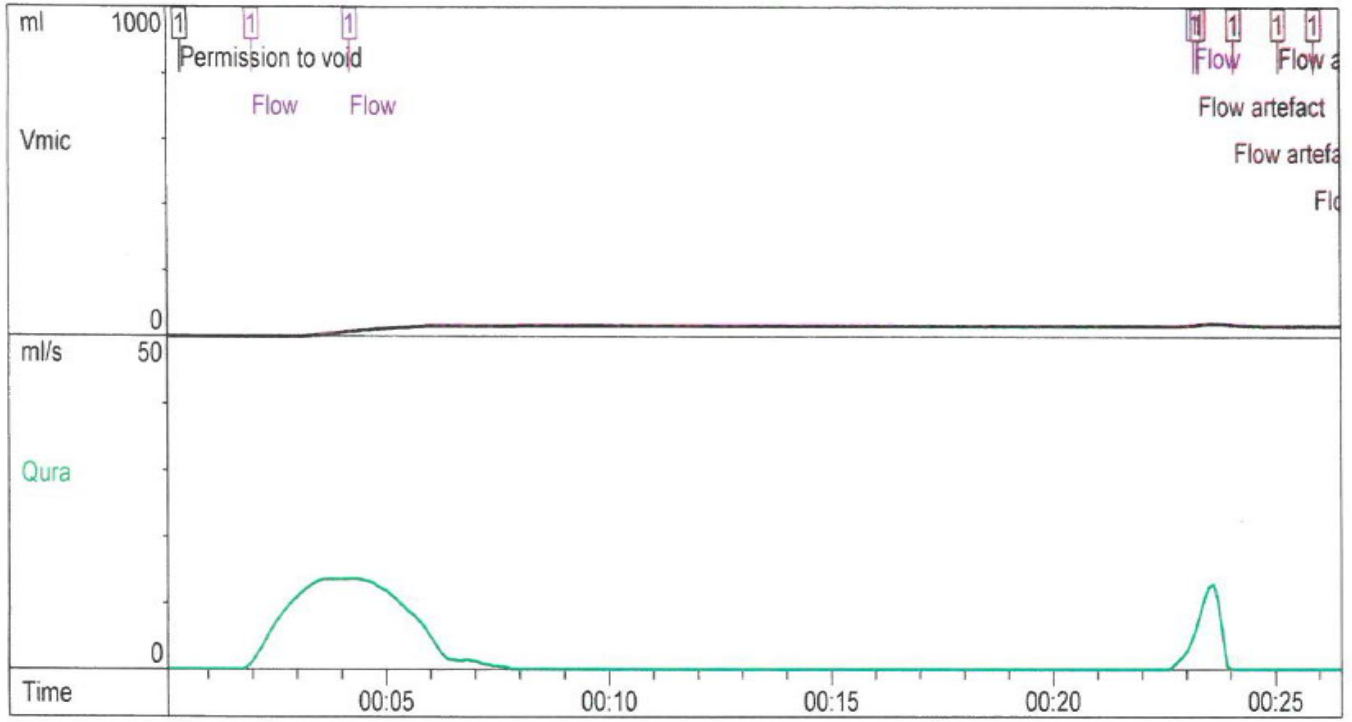
UL urinveier med resturinbestemmelse og rektal diameter

- Fortykket blærevegg?
- Hydroureter?
- Blæredivertikler?
- Rektal diameter over 3 cm?

4-timers miksjonsobservasjon

- Barn som ikke har evne til å gjennomføre flowmetri, som regel under 5 år
- Direkte etter miksjon bestemmelse av resturin
- Patologisk hvis ikke en totaltømming i løpet av 4 timer

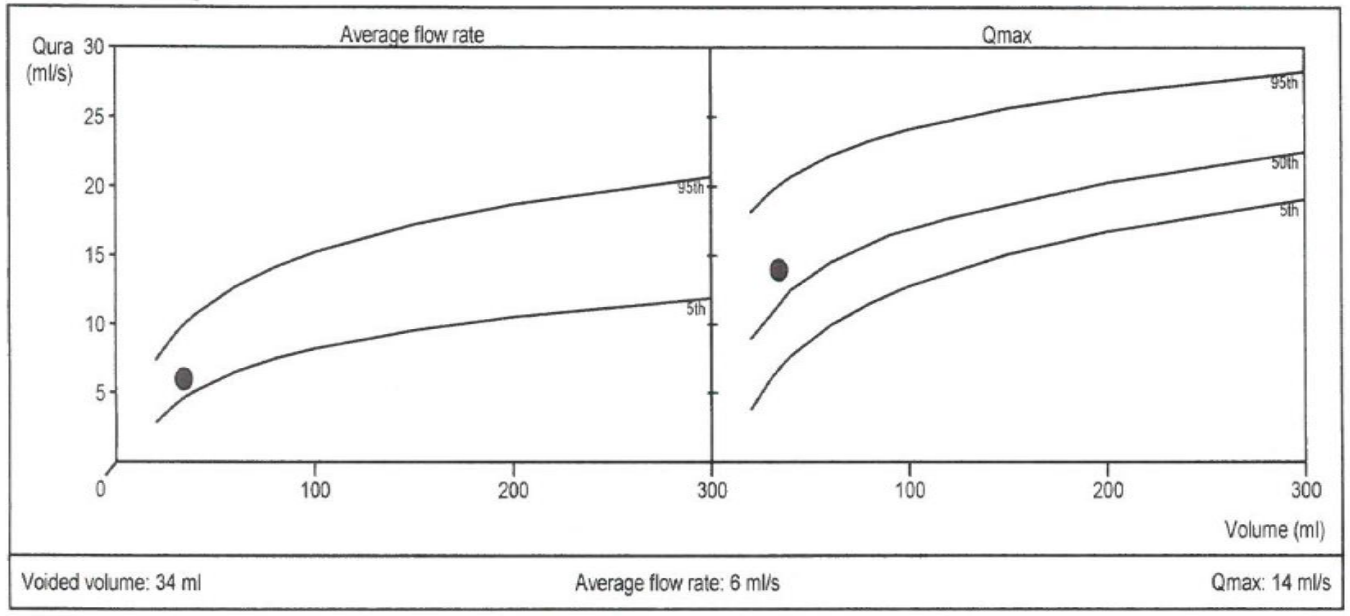




Uroflow Results

VOID	14 / 30 / 39	
Qmax	13,7	ml/s
Time to Qmax	2	s
Voided volume	34	ml
Flow time	7	s
Voiding time	22	s
Hesitancy	2	s
Average flow rate	6,2	ml/s
Corrected Qmax	6	sqrt
Residual urine	39	ml
Miction index	-1,39	ml/s

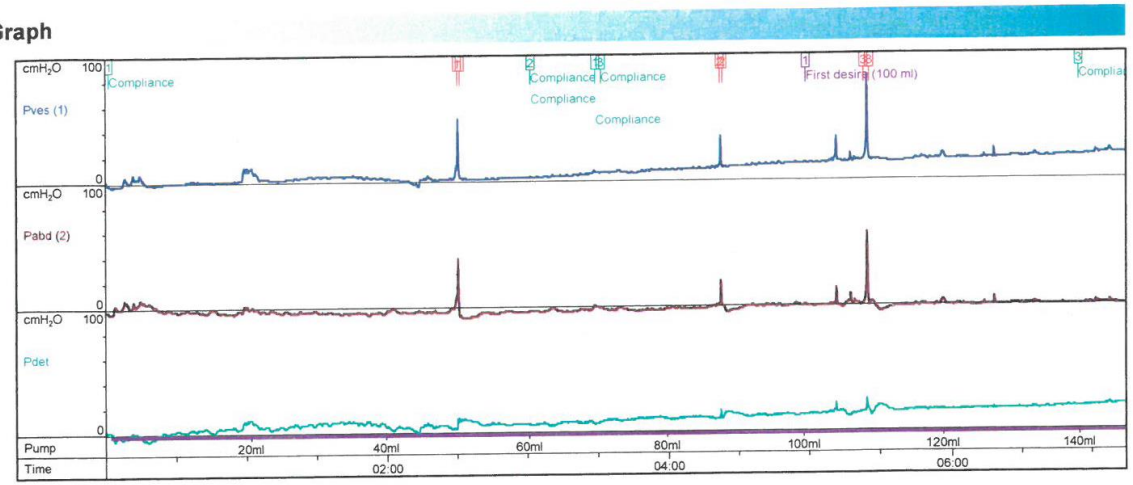
Miskolc nomogram



Diag

- Tryk
- Lekk
  und

Graph



Cystometry Results

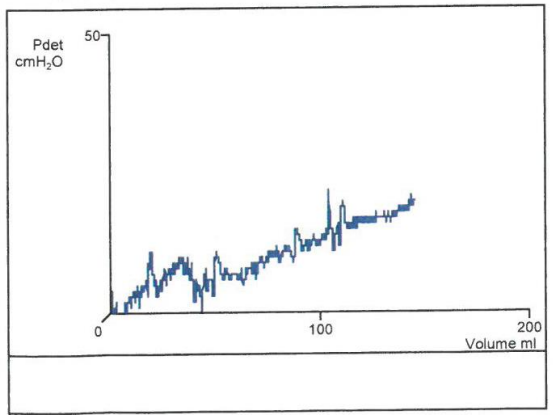
Infused volume	146 ml
Volume lost through leakage	0 ml
Bladder filling	146 ml
Maximal bladder capacity	146 ml

Sensation results

Sensation	Bladder filling ml	Pves cmH_2O	Pdet cmH_2O
First desire	100	14	13

Compliance results (linear regression)

Filling cystometrogram



#	Pves ml/cmH_2O	Pdet ml/cmH_2O	Begin ml	End ml
1	23,2	9,4	1	69
2	N/A	-	60	60
3	5,5	7,3	70	139

Oppsummering

- «Not like open neural tube defect»
- Viktig å følge opp regelmessig
- "step up" tilnærming fra minst invasive undersøkelser først til mest invasive undersøkelser på indikasjon
- Spesifikke risikofaktorer for inkontinens og RIK

Takk for oppmerksomheten!