VULVACANCER

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Vad är vulvacancer?

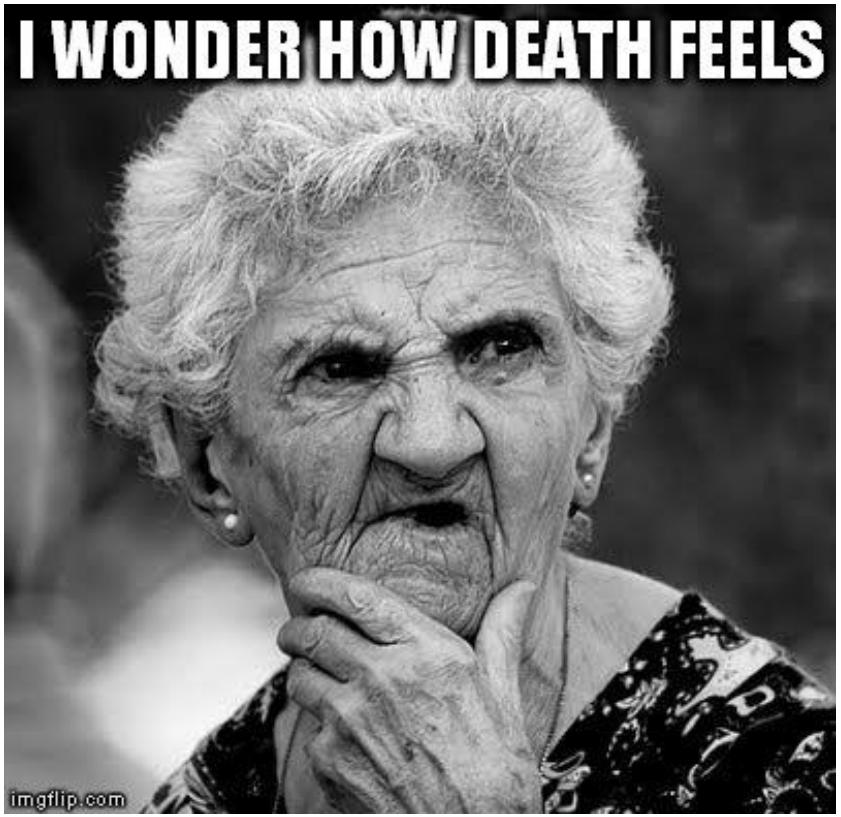
Vanligast histologisk typ:

- Skivepitelcancer 80-90%

Ytterligare histologiska typer:

- Bartholinicancer (Adeno- eller skivepitel)
- Malignt melanom
- Invasiv morbus Paget (Adeno)
- Sarkom
- Metastaser

Vem får vulvacancer?

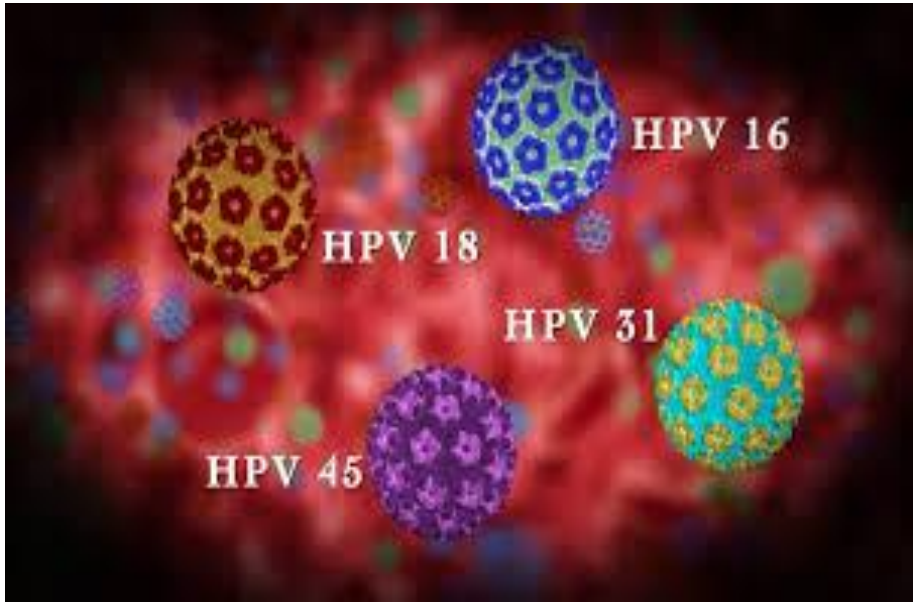


Medelålder:
70 – 75 år

HPV-relaterad?

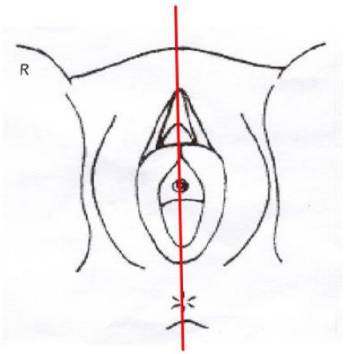
Invasiv cancer: ca 40% HPV-high-risk-positiv

VIN (dysplasi): ca 90% HPV-high-risk-positiv



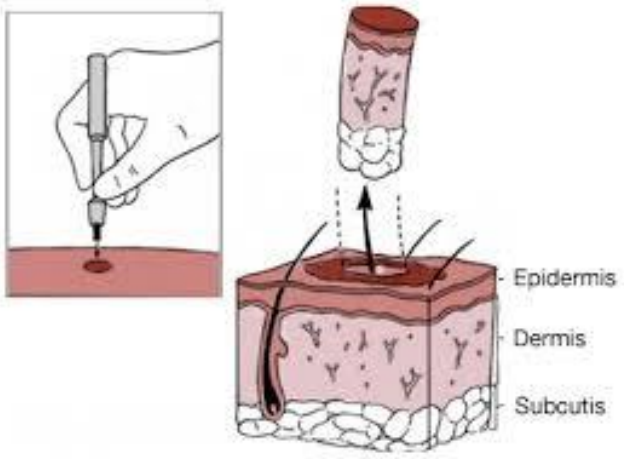
Riskfaktorer

- Ålder
- HPV-relaterad:
  - Immunsuppression
  - Rökning
- Icke-HPV-relaterad:
  - Lichen
- Status efter vulvacancer



Diagnos och utredning

- gynekologisk undersökning (lichen? VIN?)
- Biopsi (INTE cytologi i vulva!)
- Cytologi portio
- Palpation ljumskar



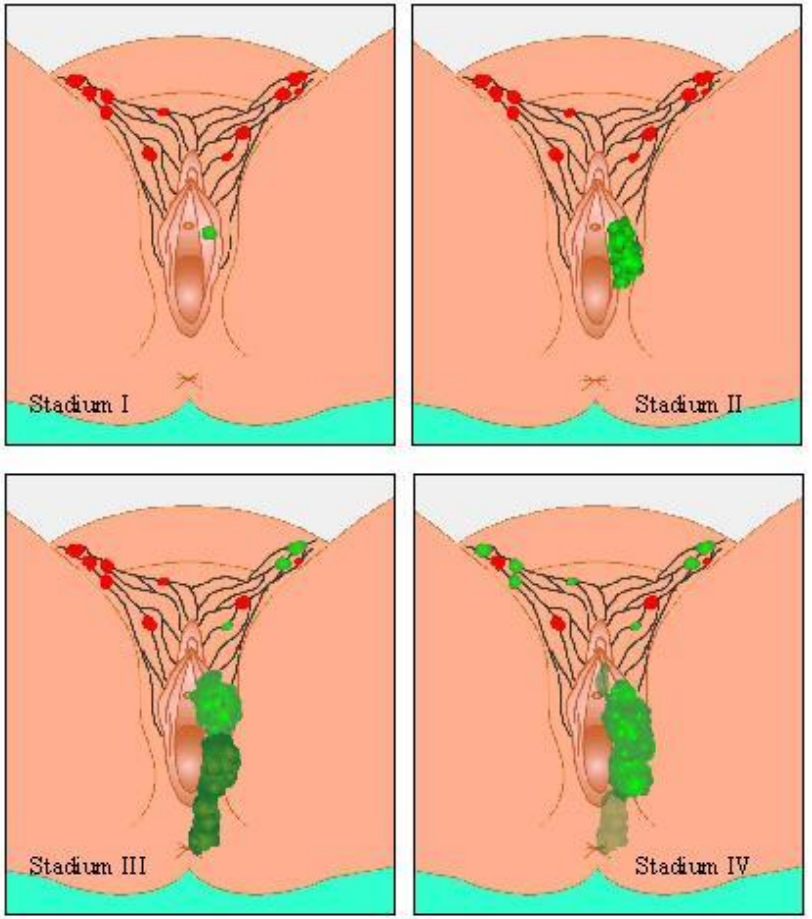
Bilddiagnostik

- Tidigt stadium (FIGO I):
  - Ultraljud ljumskar eller MR bäcken (inguinala llg-metastaser)

- Avancerat stadium (FIGO II-IV):
  - CT thorax buk
  - MR bäcken
  - PET CT???

Stadieindelning

- TNM klassifikation
- FIGO klassifkation



TNM klassifikation

Clinical *Extent of disease before any treatment*	Stage Category Definitions	Pathologic *Extent of disease during and from surgery*
❒ y clinical– staging completed after neoadjuvant therapy but before subsequent surgery	**Tumor Size:** _____ **Laterality:** ☐ left ☐ right ☐ bilateral	❒ y pathologic – staging completed after neoadjuvant therapy AND subsequent surgery

Primary Tumor (T)

TNM Category	FIGO Stage		Clinical	Pathologic
❑ TX		Primary tumor cannot be assessed	❑	❑
❑ T0		No evidence of primary tumor	❑	❑
❑ Tis	*	Carcinoma *in situ* (preinvasive carcinoma)	❑	❑
❑ T1a	IA	Lesions ≤ 2 cm in size, confined to the vulva or perineum and with stromal invasion ≤ 1.0 mm**	❑	❑
❑ T1b	IB	Lesions >2 cm in size **or** any size with stromal invasion >1.0 mm, confined to the vulva or perineum	❑	❑
❑ T2***	II	Tumor of any size with extension to adjacent perineal structures (Lower/distal 1/3 urethra, lower/distal 1/3 vagina, anal involvement)	❑	❑
❑ T3****	IVA	Tumor of any size with extension to any of the following: upper/proximal 2/3 of urethra, upper/proximal 2/3 vagina, bladder mucosa, rectal mucosa, or fixed to pelvic bone,	❑	❑

* FIGO staging no longer includes Stage 0 (Tis).

** The depth of invasion is defined as the measurement of the tumor from the epithelial-stromal junction of the adjacent most superficial dermal papilla to the deepest point of invasion.

*** FIGO uses the classification T2/T3. This is defined as T2 in TNM.

**** FIGO uses the classification T4. This is defined as T3 in TNM.

Regional Lymph Nodes (N)

TNM Category	FIGO Stage		Clinical	Pathologic
❑ NX		Regional lymph nodes cannot be assessed	❑	❑
❑ N0		No regional lymph node metastasis	❑	❑
❑ N1		One or two regional lymph node with the following features	❑	❑
❑ N1a	IIIA	One or two lymph node metastasis each 5 mm or less	❑	❑
❑ N1b	IIIA	One lymph node metastases 5 mm or greater	❑	❑
❑ N2	IIIB	Regional lymph node metastasis with the following features:	❑	❑
❑ N2a	IIIB	Three or more lymph node metastases each less than 5 mm	❑	❑
❑ N2b	IIIB	Two or more lymph node metastases 5 mm or greater	❑	❑
❑ N2c	IIIC	Lymph node metastasis with extracapsular spread	❑	❑
❑ N3	IVA	Fixed or ulcerated regional lymph node metastasis	❑	❑

An effort should be made to describe the site and laterality of lymph node metastases.

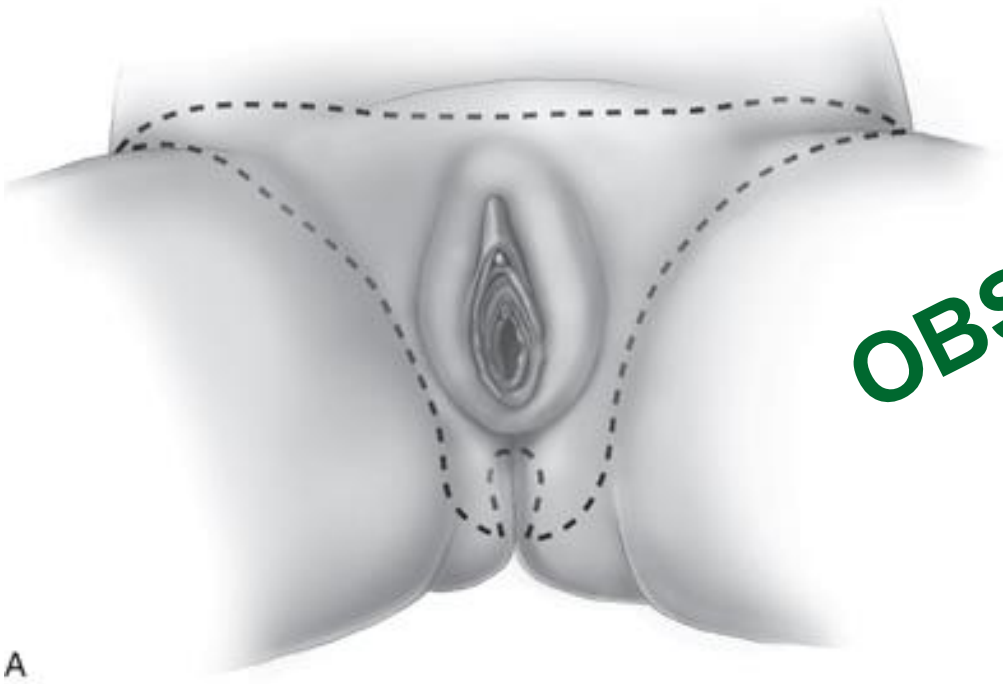
Distant Metastasis (M)

TNM Category	FIGO Stage		Clinical	Pathologic
❑ M0		No distant metastasis (no pathologic M0; use clinical M to complete stage group)	❑	
❑ M1	IVB	Distant metastasis (including pelvic lymph node metastasis)		❑

Hur behandlar vi vulvacancer?

Kirurgi!

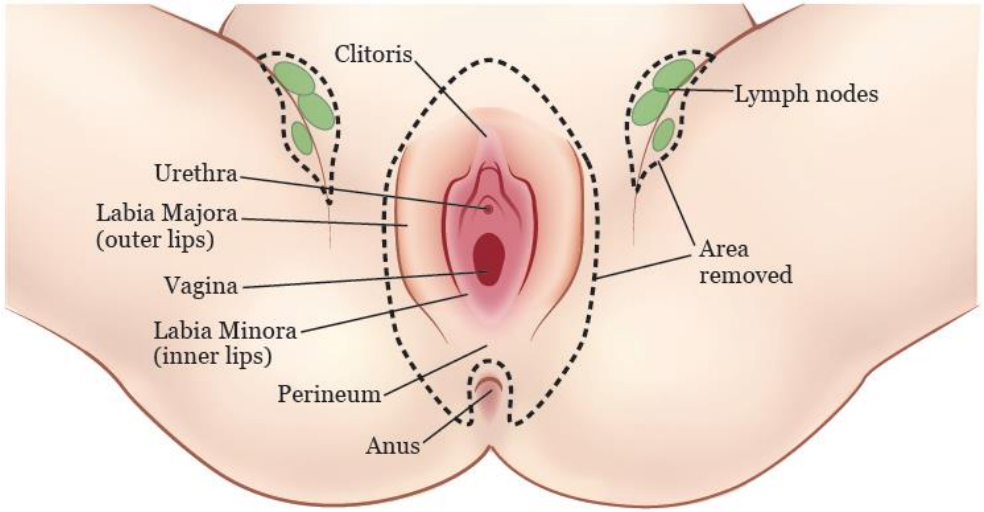
Kirurgisk behandling: Butterfly incision



A

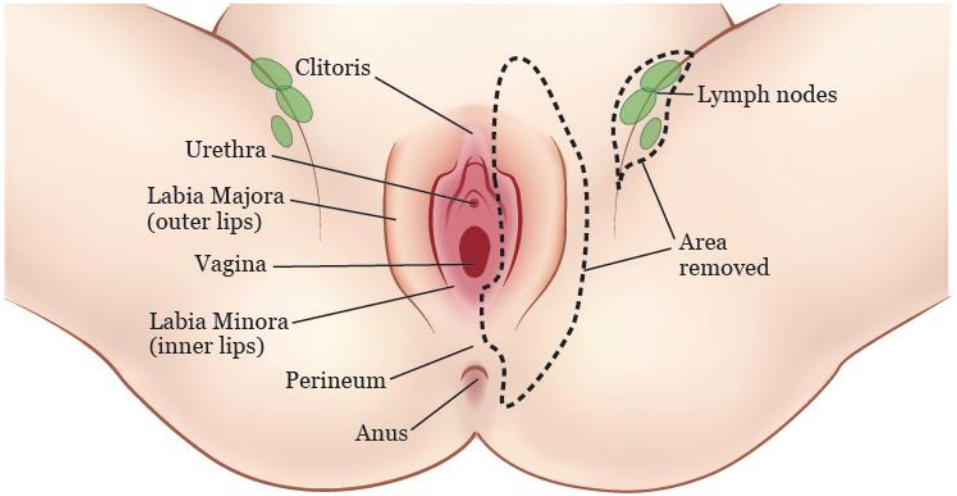
OBSOLET

Kirurgisk behandling

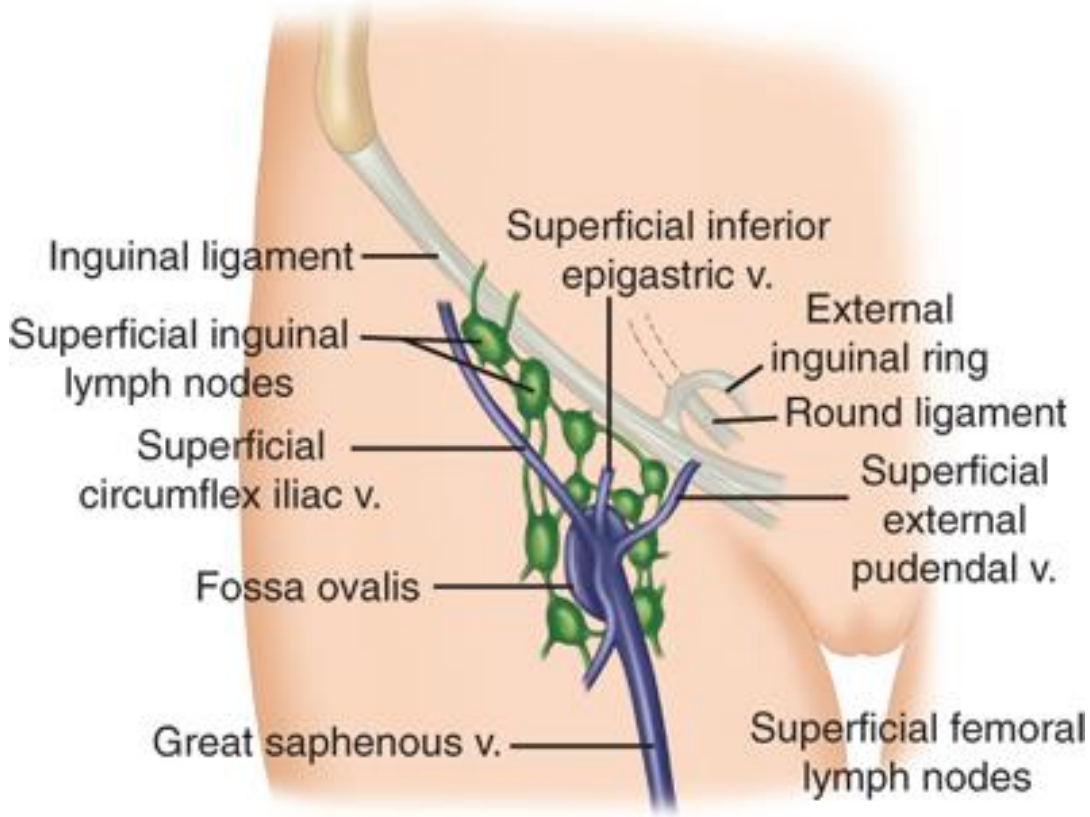


Vulvektomi

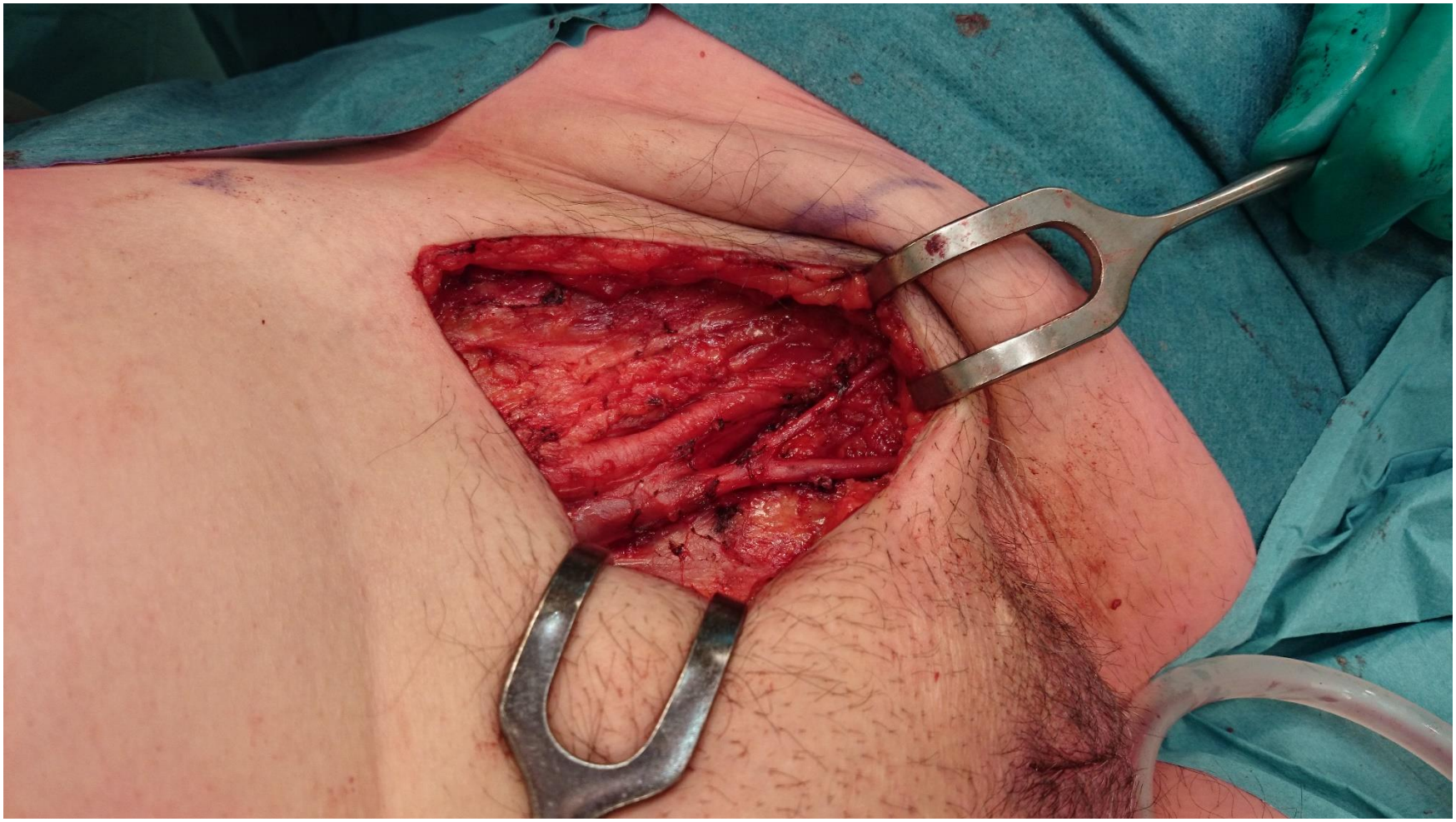
Vulvaresektion



Anatomi – ljumskar



Kirurgisk behandling: Lymfkörtelutrymning



Kirurgisk behandling: Sentinel node teknik

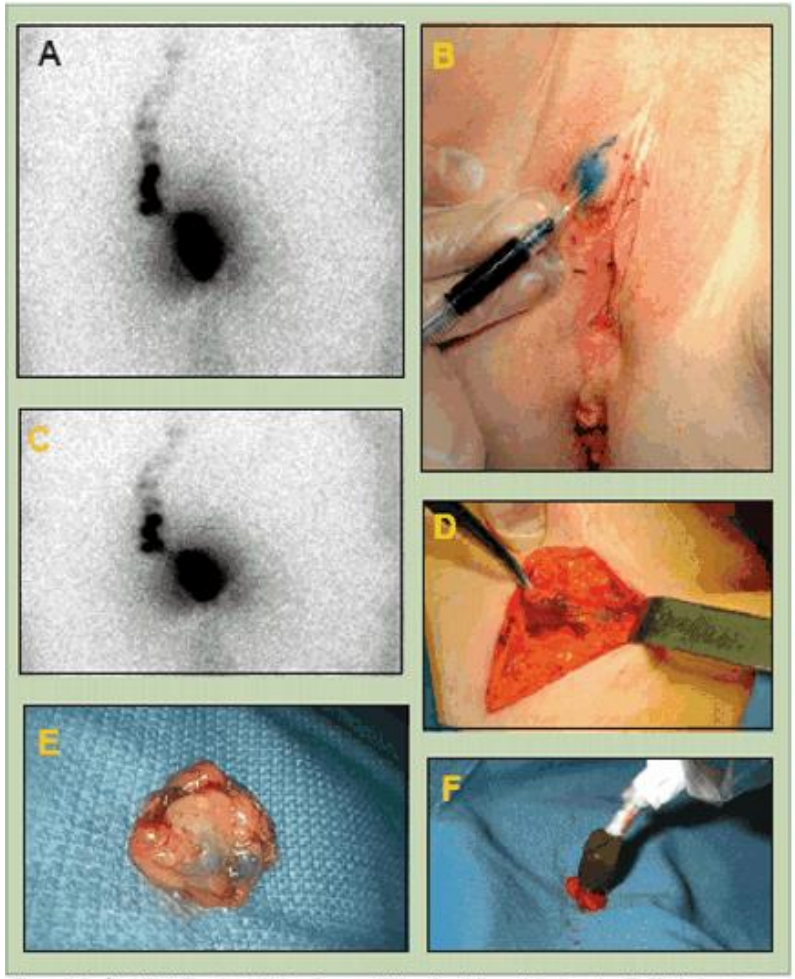
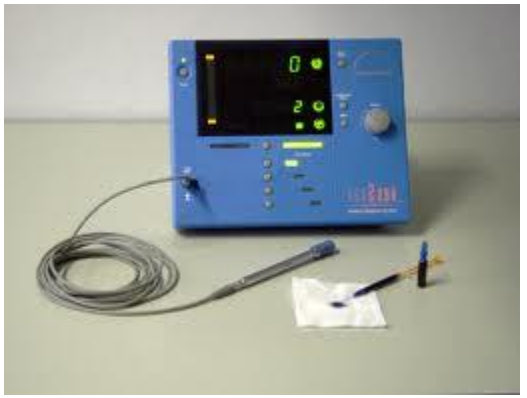
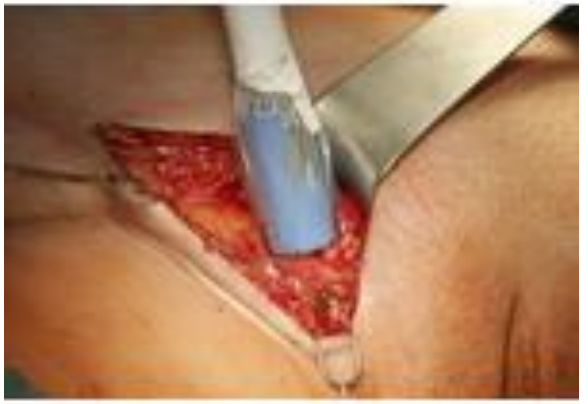


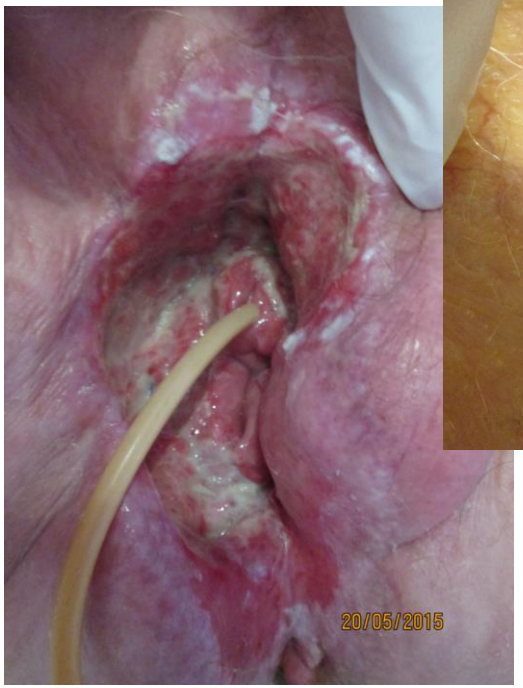
Figure 1: Sentinel Node Mapping in Vulvar Cancer—(A) Lymphoscintigram following vulvar Tc-99 injection showing right-sided groin uptake. (B) Blue dye injection at the periphery of the tumor. Note the onset of lymphatic tracking under the skin. (C) Right groin exploration with the handheld probe. (D) Right groin exploration searching for the blue node. Note the blue lymphatics entering the sentinel node. (E) Two sentinel nodes removed en bloc with fatty tissue. (F) Extracorporeal count performed with the handheld probe to help determine which of the two sentinel nodes is the primary, and to confirm that the hot sentinel node detected in the groin is really the one that has been removed.

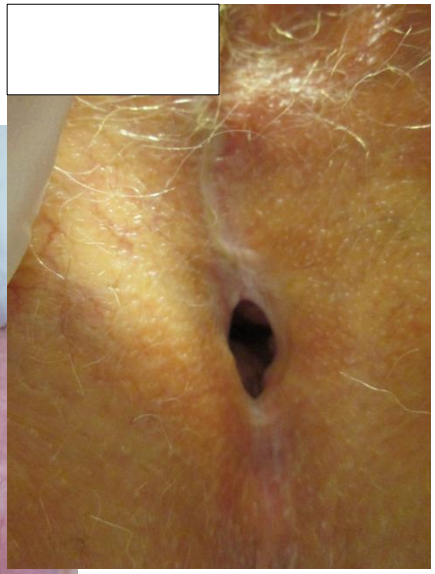


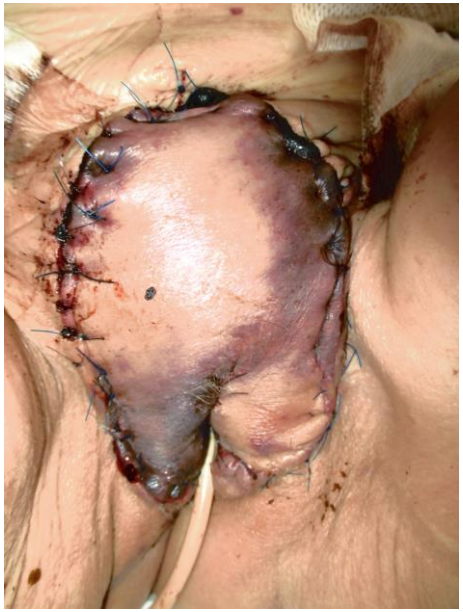


Typiska komplikationer efter kirurgi - vulva

- Sårinfektion
- Sårruptur
- Dehiscens lambå
- Nekros lambå







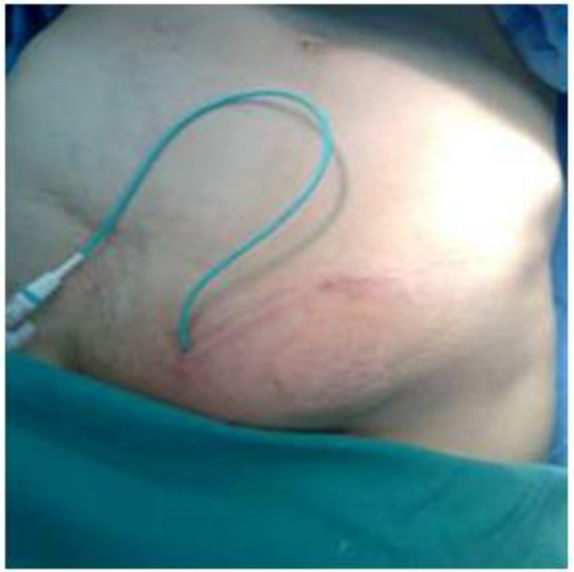
- <u>Sent:</u>

  stenoseringar, urininkontinens,

  samlagsbesvär, skavproblem

Typiska komplikationer efter kirurgi - ljumskar

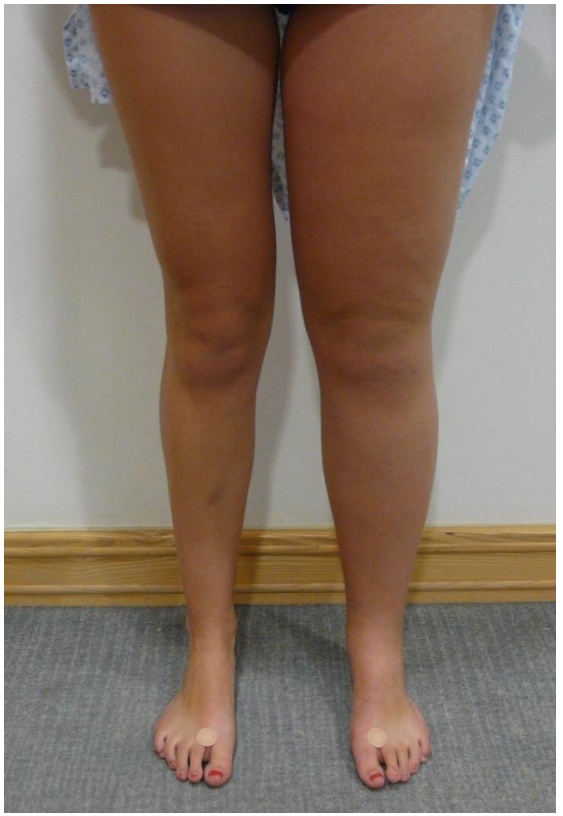
- Sårinfektion
- Sårruptur
- Lymfocele
- Hudnekros



- <u>Sent:</u>

  Recidiverande erysipelas

  Lymfödem



Annan behandling: Strålbehandling

- Primär:
  - kurativt
  - palliativt (symptomlindrande)
- Adjuvant:
  - kurativt
- Vulva:
  - adjuvant efter operation pga dålig marginal
  - primär istället för operation
- Ljumskar:
  - adjuvant efter operation vid metastaser i lymfkörtler
  - palliativ istället för kirurgi

Annan behandling: Cytostatikabehandling

- Som tillägg till strålbehandling (konkomitant radiokemoterapi)
- Primär eller adjuvant
- Enbart cytostatikabehandling enbart i palliativt syfte (dålig effekt)

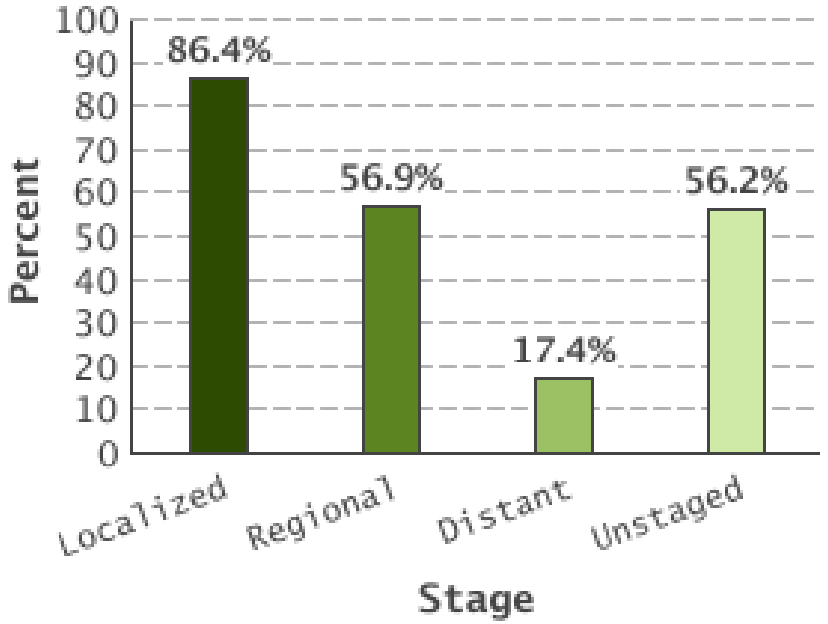
Annan behandling:

Nix

(Erlotinib?)
(Immunterapi?)

Prognos???

- Viktigaste prognosfaktorn: Lymfkörtelstatus i ljumskar
- Vanligast: lokalrecidiv (20-35%)
- Ofta mycket sena recidiv (”de-novo”-tumörer?)
- God prognos vid lokalrecidiv
- Dålig prognos vid lymfkörtelrecidiv
- Mycket dålig prognos vid distant recidiv (fjärrmetastaser)



5-års relativ överlevnad, SEER-report, 2007-2013, USA

https://seer.cancer.gov/statfacts/html/vulva.html

Överlevnad GROINSS-studie

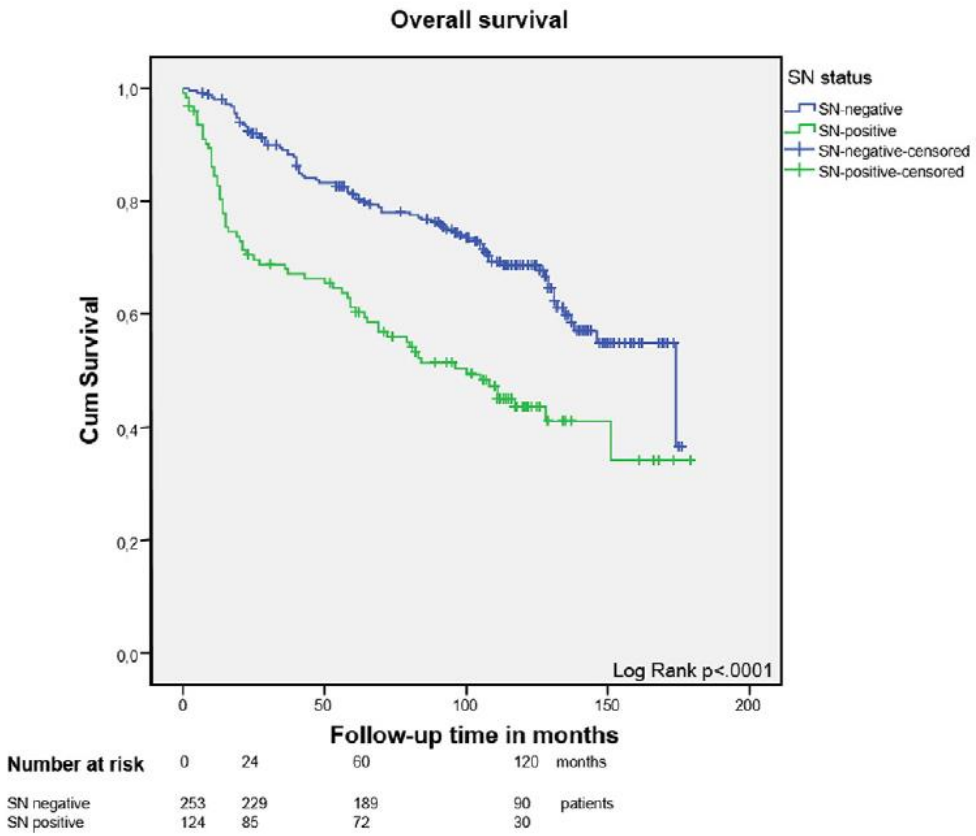


Fig. 4. Overall survival for SN-negative and SN-positive patients.

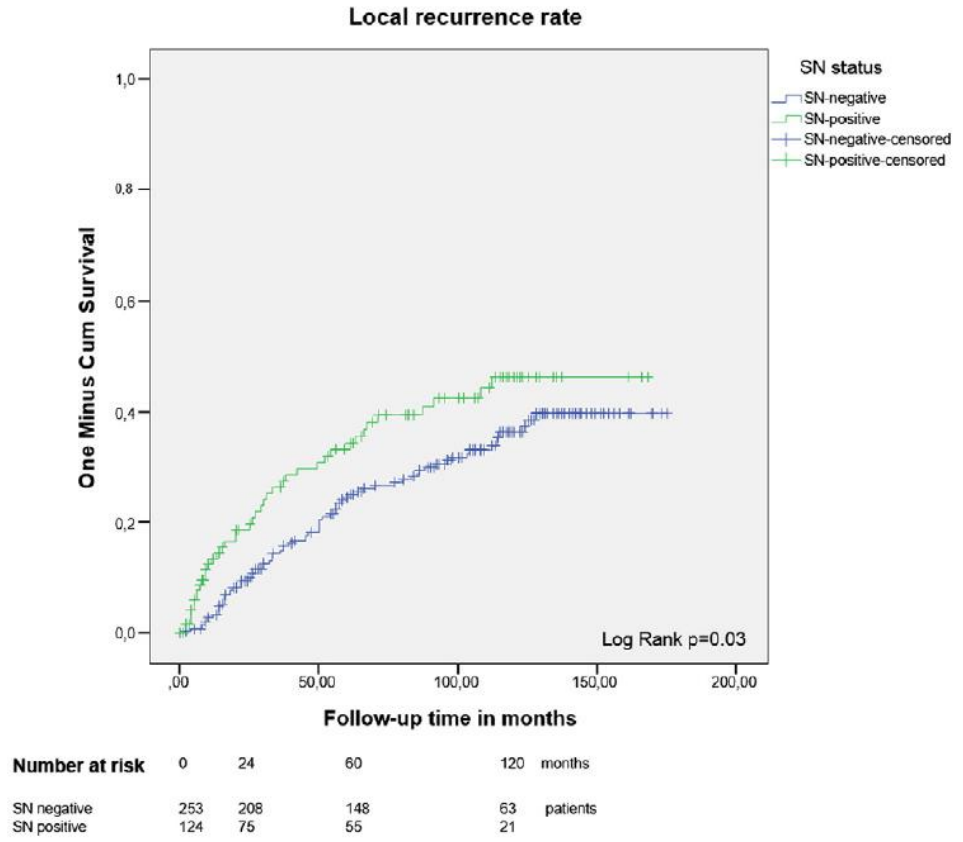


Fig. 2. Local recurrence rate for SN-negative and SN-positive patients.

te Grootenhuis N c., van der Zee A g. j., van Doorn H c., van der Velden J, Vergote I, Zanagnolo V, et al. Sentinel nodes in vulvar cancer: Long-term follow-up of the GROningen INternational Study on Sentinel nodes in Vulvar cancer (GROINSS-V) I. Gynecol Oncol. 2016 Jan 1;140:8–14.

Nivåstrukturering för behandling av vulvacancer

- 4 centra i Sverige: Karolinska Stockholm, Sahlgrenska Göteborg, Linköpings universitetssjukhus, Lund universitetssjukhus
- Utomlänspatienter från hela landet (ca 10-15 per år)
- En gång per vecka nationell multidisciplinär konferens nMDK
- Nationellt vårdprogramm, standardiserat vårdflöde SVF, kvalitetsuppföljningskrav

Premaligna vulvaförändringar

- Lichen
- VIN:
  - dVIN
  - uVIN / HSIL
- LSIL (kondylom, ingen dysplasi, ingen precancerös förändring)
- Morbus paget (icke invasiv, primär eller sekundär)
- Melanoma in situ

Premaligna vulvaförändringar: Vulvadysplasi

Kaufmann, 1965:

Queyrat's erythroplasia

Bowenoid carcinoma (Mb Bowen)

Carcinoma simplex

Premaligna vulvaförändringar: Vulvadysplasi

ISSVD (International Society for the Study of Vulvar Disease),1976:

Vulvar atypia

Carcinoma in situ

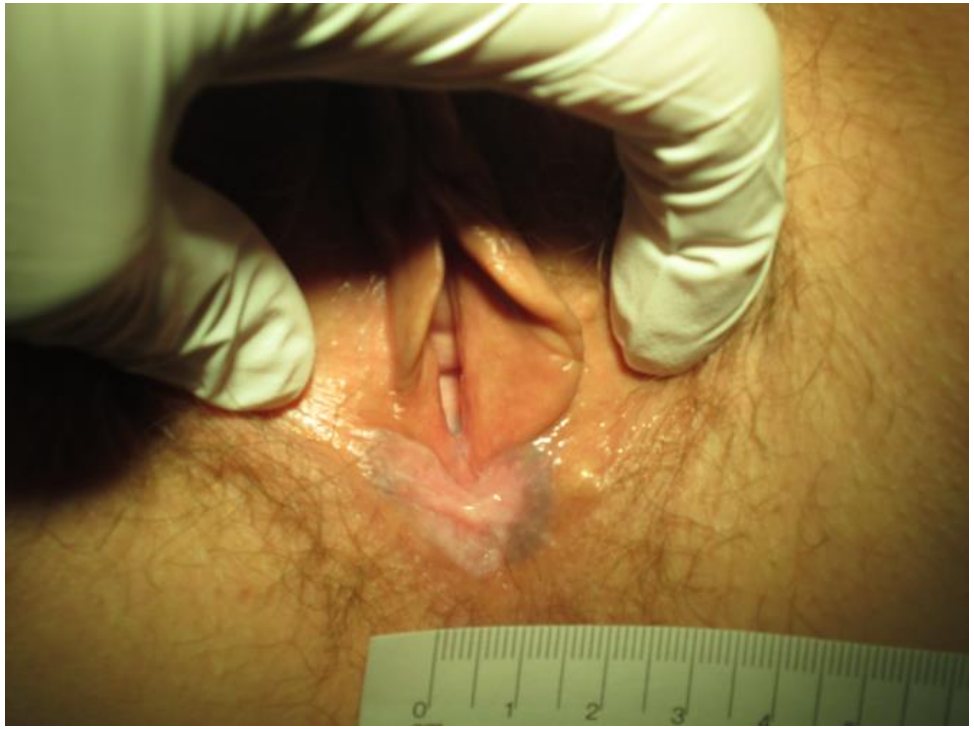
Premaligna vulvaförändringar: Vulvadysplasi

ISSVD / WHO, 1986:

~~VIN I~~

~~VIN II~~

VIN III



Premaligna vulvaförändringar: Vulvadysplasi

LAST (Lower Anogenital Squamous Terminology) 2012
ISSVD 2015:

Usual VIN (uVIN) / HSIL:

- Warty
- Basaloid
- Mixed

HPV-relaterad

Differentiated VIN (dVIN)

icke-HPV-relaterad

Alltid „High grade“ (VIN II-III / HSIL)

„VIN I“ / LSIL = ingen dysplasi