

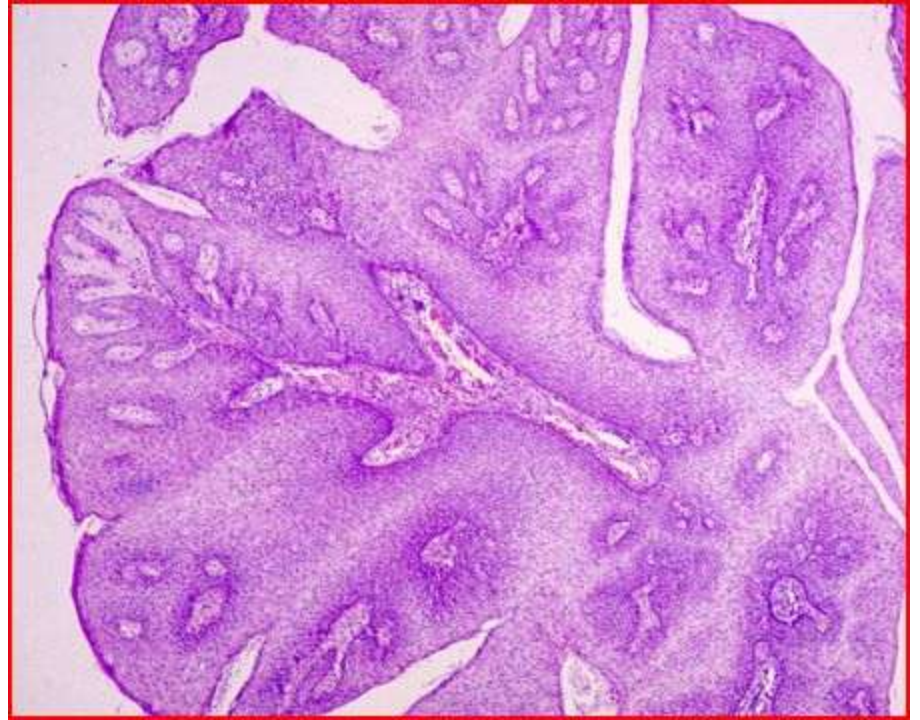
Juvenile Recurrent Respiratory Papillomatosis

Definition: JRRP

- Juvenile
 - Mean age at diagnosis 3years
 - More aggressive than adult disease
- Recurrent
 - Average lifetime procedures = 21
- Respiratory
 - Usually larynx
 - If extends below larynx tends to be younger
- Papillomatosis
 - With rare dysplasia and progression to carcinoma

Pathology

- Papilloma virus induces abnormal differentiation of epithelium, resulting in relative hyperplasia of the parabasal layers and papillomatous formation



Aetiology



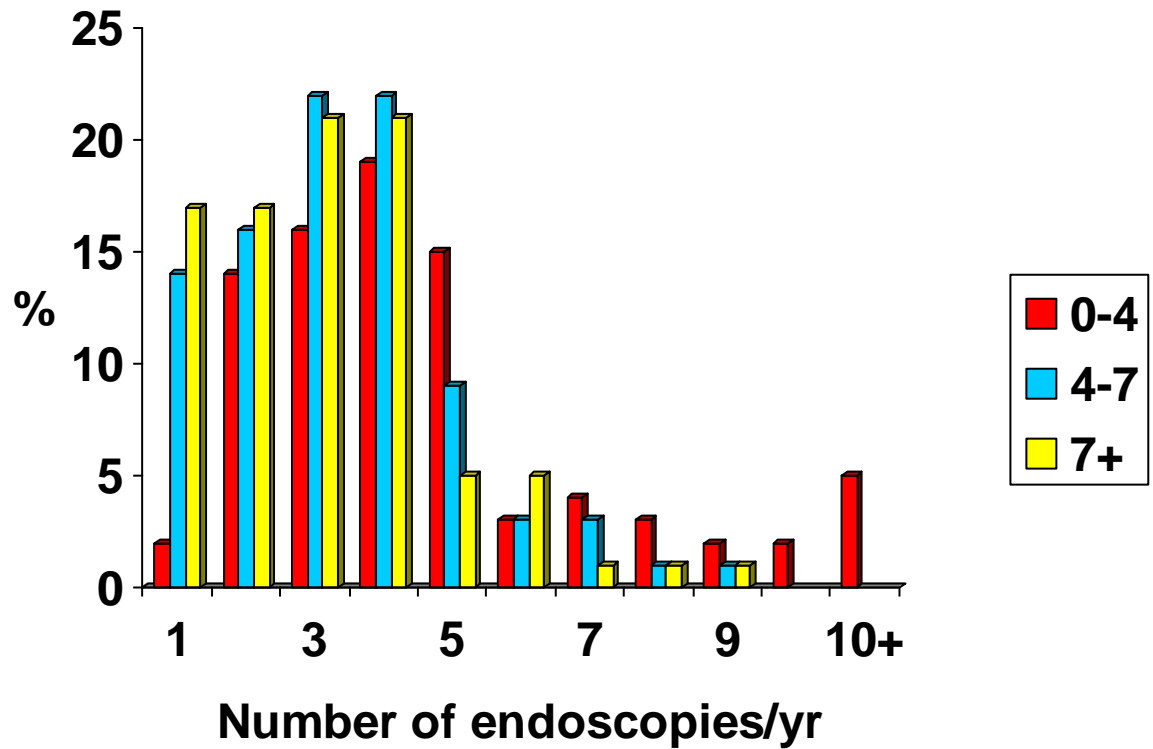
HPV types

- -
 -
 -
- 6
- Benign genital warts/JRRP
- 11
- In JRRP 11 associated with more severe disease
- -
 -
- 16
- Cervical cancer
- 18

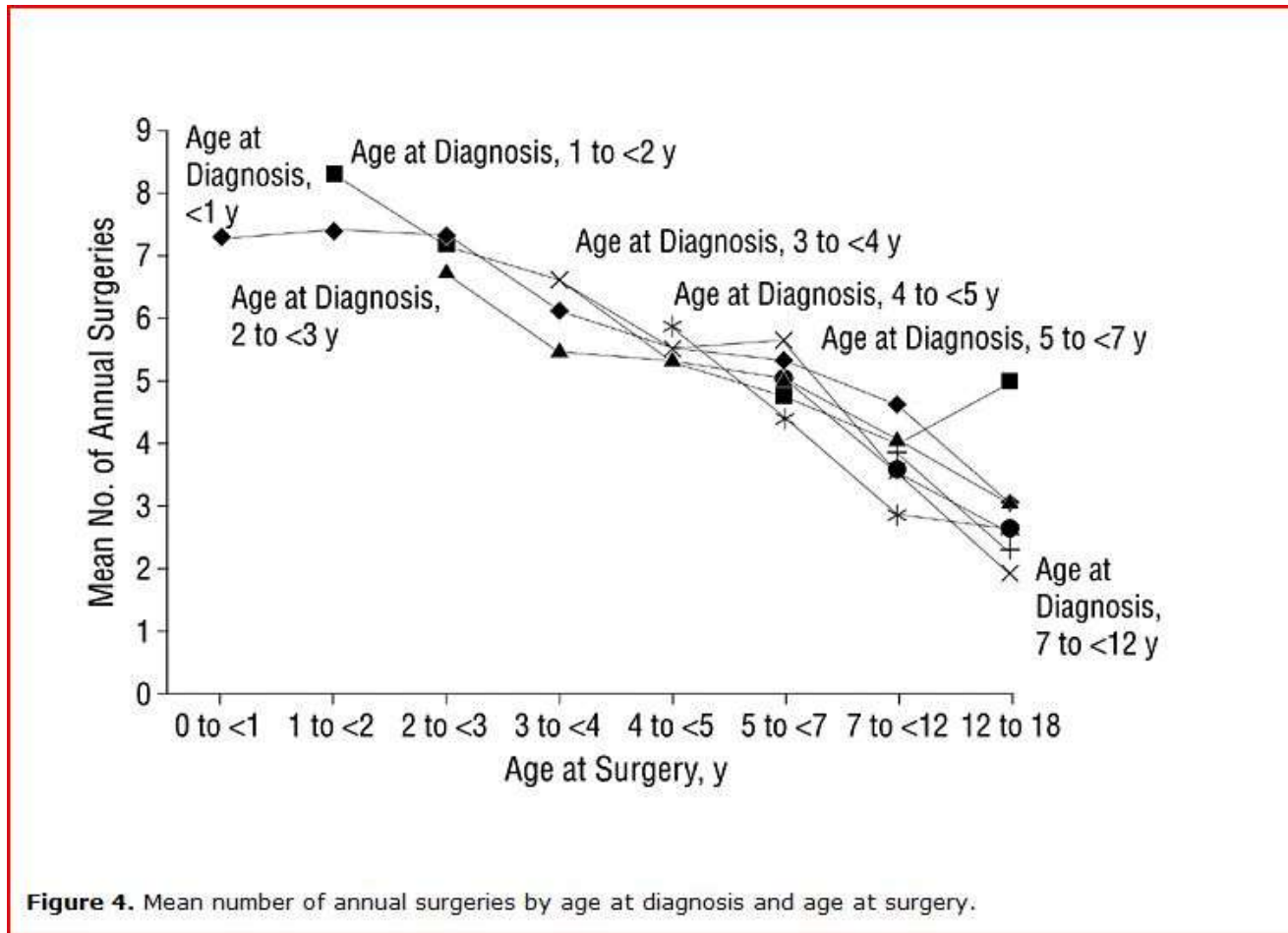
HPV facts

- JRRP: 50% have family history of genital tract HPV infection
- Patients delivered by Cesarean section are not immune to developing RRP
 - ? infection transplacental or postnatal
 -
- HPV common in general population therefore overall risk of JRRP is low
- HPV viral DNA and capsid antigen found in macroscopically normal tissues. (even in remission)
- A latent form of virus would explain recurrence and spread to previously uninvolved sites.
- Surgery alone is not the answer!

Age at diagnosis related to prognosis



Gradual tendency to improve with age



Clinical features

- *Progressive* hoarseness
- Dyspnoea
- Noisy breathing
- Stridor

Endoscopy

- Fibreoptic exam in the office
 - Should differentiate vocal cord nodules from papillomatosis
 - Should exclude massive supraglottic lesion so that anaesthesia is warned
- Formal endoscopy required for accurate diagnosis and treatment

Radiology

- Baseline chest xray
- MRI once disease spreads to trachea to exclude parenchymal disease

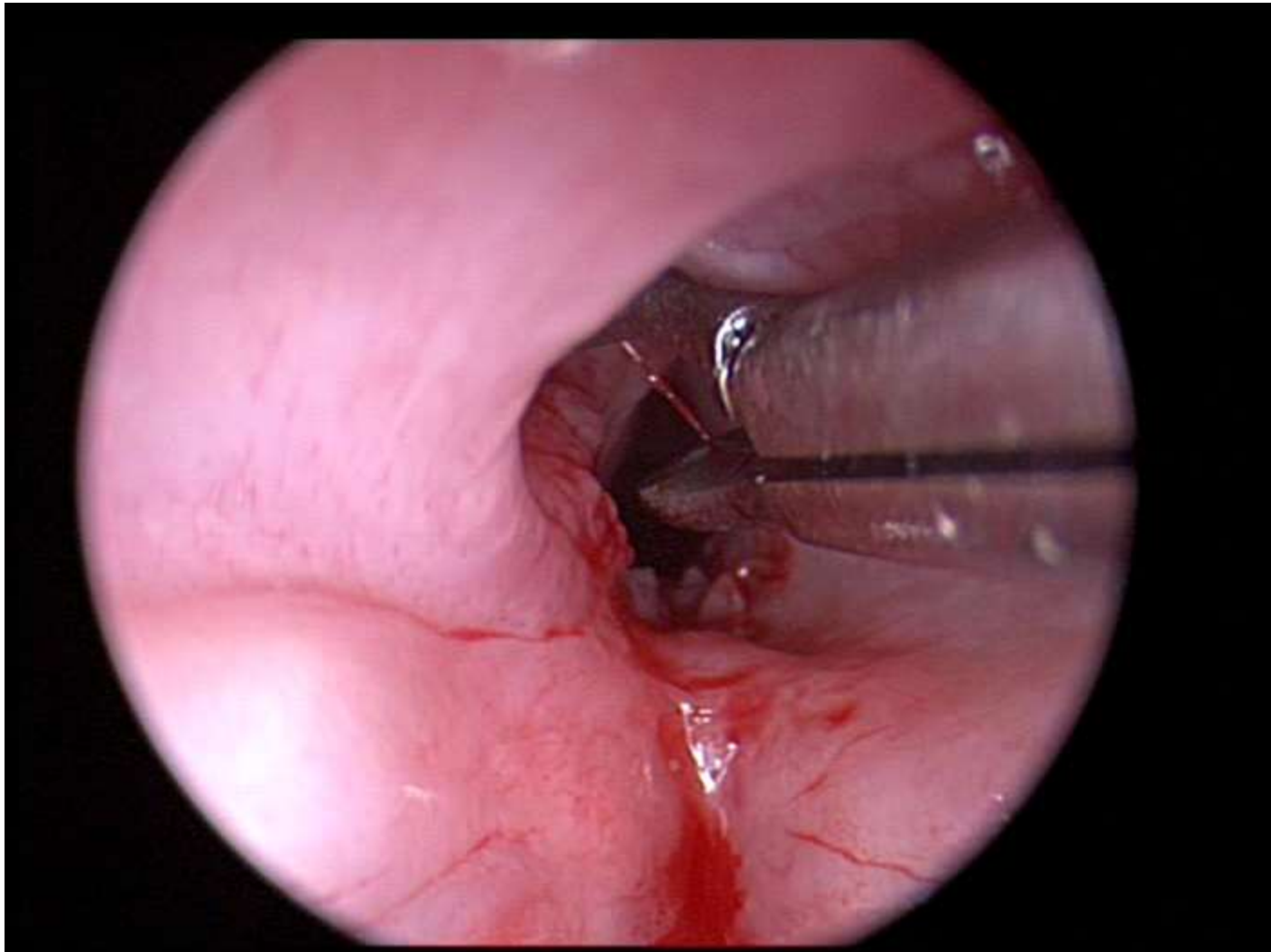
MLB

- Confirm diagnosis
 - Biopsy
 - Viral Typing
- Record anatomical sites and spread
 - Photo documentation
 - Derkay score

Surgical Treatment

- Cold Steel resection with cup forceps
- CO₂ or KTP Laser
- Microdebrider
- Tracheostomy

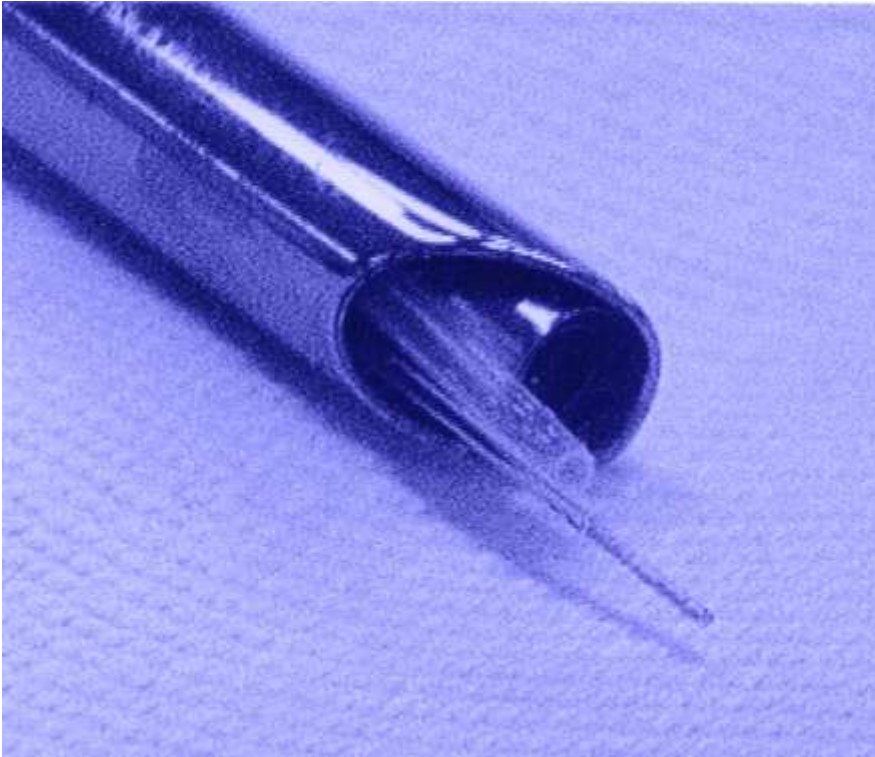
Cold steel resection



Microdebrider



Laser



Tracheostomy

- About 10% require tracheostomy
- Children with tracheostomy tend to have severe and extensive disease
- Not clear if trach is needed in severe disease
OR Trach causes spread
- No suggestion that disease progresses after tracheostomy is placed

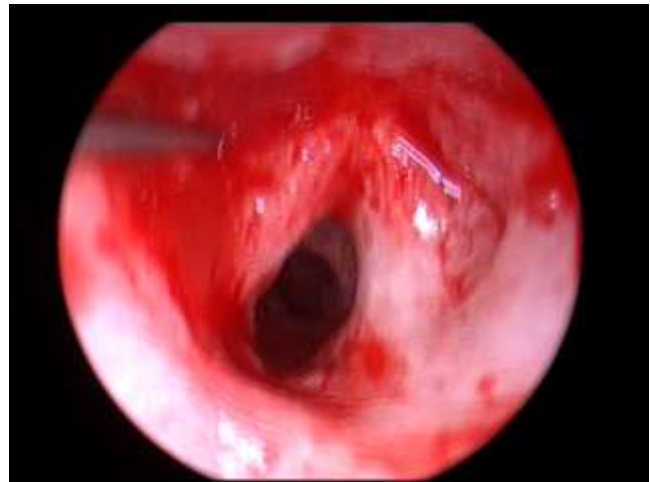
Adjunctive treatments

- Interferon

- Cidofovir

 - 5mg/ml

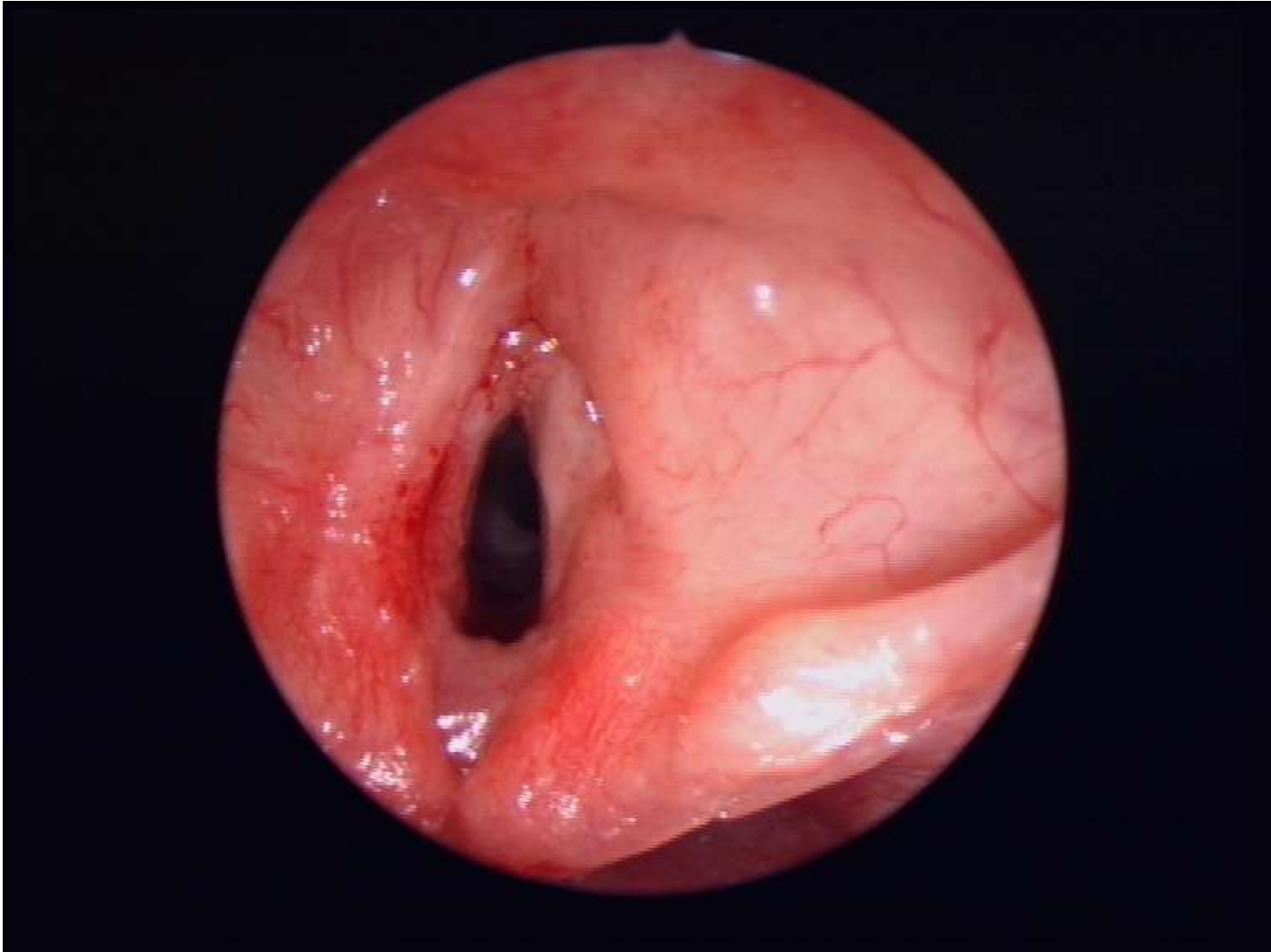
 - 3ml = 15mg



Ist case with Cidofovir



Recent case



Cidofovir

- Does it work?
- Is it safe?

Severe disease

- Endoscopy every 2-4 weeks
- Extensive tracheal spread
- Evidence of lung parenchymal spread

Time: 12:34:28

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Se: 5

x 1.4



Scan:

SP:

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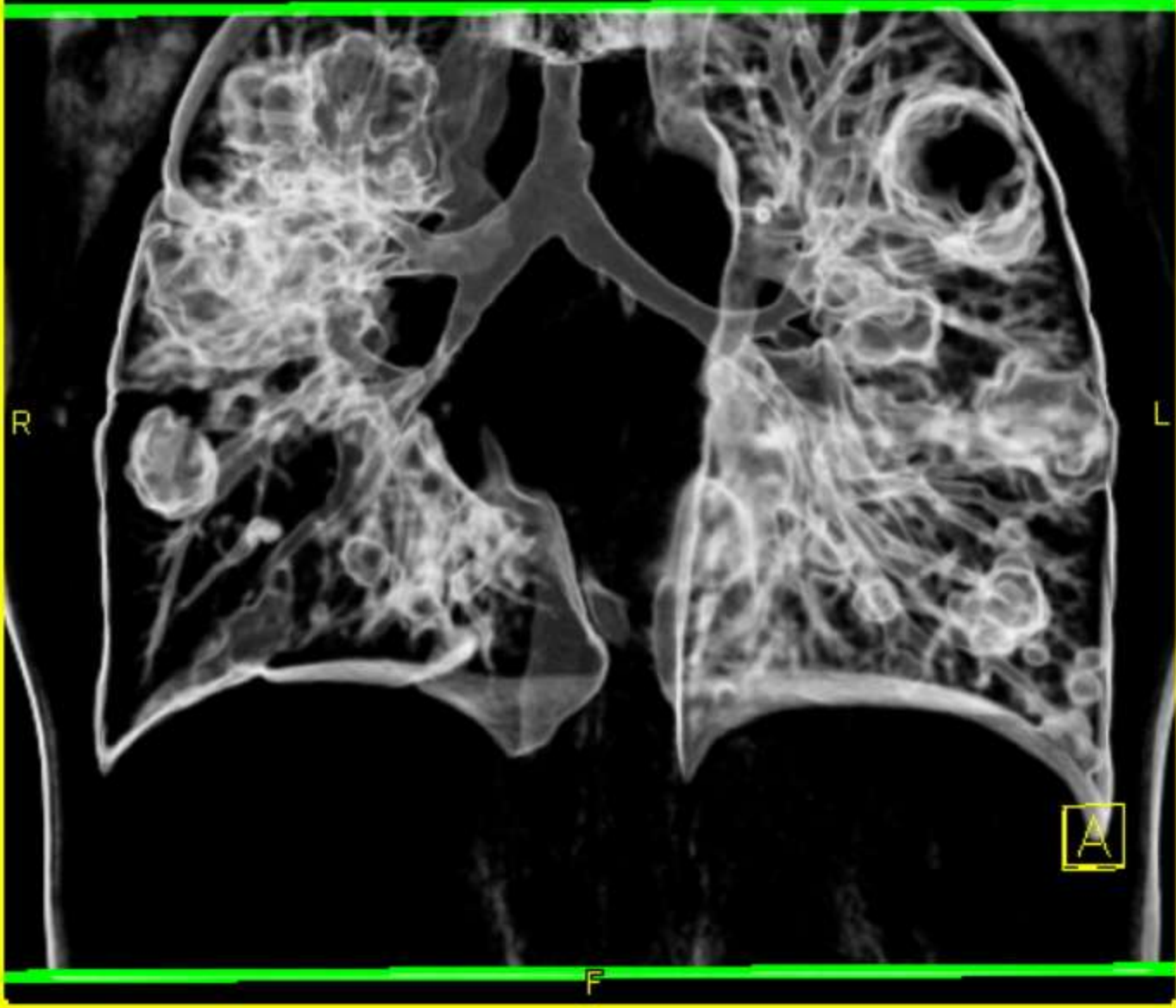
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Rows: 512

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Severe disease

- IV Cidofovir
- Instilled cidofovir
- Interferon
- Regular ciproxin

- Regular MRI to assess progression/ca
- Regular biopsy

Core technique



Future

- HPV Vaccination



- Gardasil Quadravalent Vaccine 6/11/16/18

- » (Vaccine to L1 Capsid antigen, intranuclear so not accessible, therefore not therapeutic)

- Cervarix 16/18 (75% cervical ca) - NHS!!!!

- ? Changes in HPV type prevalence

- E6 mRNA produce proteins needed for replication and would be accessible for suitable vaccine

