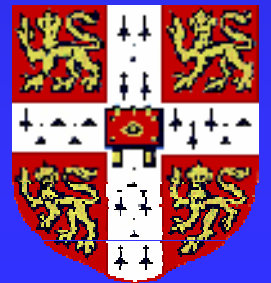


Disease Stage

“What is going to happen to me?”

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Recent Diagnosis

- How will the condition progress?
- Investigations / disease monitoring
- Controlling symptoms
- Controlling disease / treatment complications
- 'Short' term outcomes

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Disease Progression

- Depends on actual condition and severity
 - ◆ constitutional vs. organ-specific symptoms
 - ◆ chronic, low grade “grumbling” symptoms
 - ◆ bouts of symptoms interspersed with feeling well
 - ◆ continuing deterioration
 - ◆ induction & maintenance of remission
 - ◆ longer term – cycles of remission / relapse

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Investigations & Monitoring (1)

- Aims of management
 - ◆ control symptoms
 - ◆ control underlying disease
 - ◆ minimise treatment related complications
 - ◆ improve clinical outcome & quality of life
- Essential to monitor disease activity

Investigations & Monitoring (2)

- Often a repeat of those used to establish diagnosis
 - ◆ standardised disease activity index
 - ◆ e.g. BVAS, VDI
 - ◆ clinical symptoms & signs
 - ◆ laboratory tests – ANCA, CRP, ESR
 - ◆ clinical tests – nerve conduction studies
 - ◆ radiology – x-rays, CT scans
 - ◆ very rarely repeat biopsy, angiogram etc.

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Symptom Control (1)

- Are symptoms due to ?
 - ◆ active disease
 - ◆ disease related scarring
 - ◆ treatment-related
- Symptom control achieved by
 - ◆ treating / modifying the symptom
 - ◆ treating the underlying disease
 - ◆ reviewing need for / type of additional therapy

Symptom Control (2)

- Symptom control
 - ◆ many manifestations take months to improve
 - ◆ frequently incomplete
 - ◆ ‘constitutional’ symptoms can be troublesome

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Disease Control (1)

■ Aims

- ◆ induce & maintain remission
- ◆ minimise toxicity

■ Depends on disease severity

- ◆ steroids & immunosuppressive agent
- ◆ dose and therapy adjustment with time / response
- ◆ biological agents
- ◆ plasma exchange (plasmapheresis)

Disease Control (2)

- Drug toxicity
 - ◆ depends on degree of immunosuppression
 - ◆ most are relatively mild
 - ◆ ~ 25% of patients have severe drug related complication in first 18 months

Disease Control (3)

- Other therapies

- ◆ other preventative therapies whilst on high dose immunosuppression
 - ◆ bone protection
 - ◆ antibiotics & anti-fungal agents
 - ◆ anti-indigestion therapy

Disease Control (4)

- Ultimately transition to maintenance therapy
 - ◆ combination therapy
 - ◆ at 3-6 months
 - ◆ 12-18 months duration

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'Short' Term Outcome

- With treatment
 - ◆ ANCA associated vasculitis
 - ◆ ~ 75% in remission at 3 months
 - ◆ > 90% at 6 months
 - ◆ mortality (Wegener's granulomatosis)
 - ◆ previously 90% mortality at 2 years
 - ◆ now 10-20% at 5 years

Summary

- Frequent & regular re-assessment essential
- Treatment
 - ◆ symptom control
 - ◆ disease modifying
- Aim to induce remission & minimise complications
- Symptom resolution may be slow and incomplete

