Developments in the management of idiopathic pulmonary fibrosis

**Question 1**
In the UK what is the approximate annual incidence of IPF?
- a. 1,000
- b. 2,500
- c. 5,000
- d. 15,000
- e. 25,000

Answer: c. IPF has an incidence in the UK of around 5,000 new cases per year and the prevalence is around 15-25 in 100,000.

**Question 2**
What is the approximate 5-year survival rate for patients with IPF?
- a. 5%
- b. 20%
- c. 35%
- d. 50%
- e. 65%

Answer: b. With a median survival of 3 years and 5-year survival rate of around 20%, IPF has a prognosis worse than many cancers.

**Question 3**
Two randomised double-blind placebo-controlled studies have assessed the use of pirfenidone in adults with mild-to-moderate IPF. When the 2,403mg-dose arms of both studies were combined what was the difference in mean decline in percentage of predicted forced vital capacity (FVC) with pirfenidone compared with placebo?
- a. 1%
- b. 2.5%
- c. 5%
- d. 7.5%
- e. 10%

Answer: b. A significantly reduced mean decline in percentage of predicted FVC was seen with pirfenidone 2,403mg daily compared with placebo in one study (−8.0% vs. −12.4%, p=0.001), but not in the other (−9.0% vs. −9.6%, p=0.501). When the 2,403mg-dose arms of both studies were combined there was a significant treatment effect in favour of pirfenidone (−8.5% vs. −11.0%, p=0.005).

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Question 4

Two concurrent randomised placebo-controlled trials have assessed nintedanib 150mg twice daily in 1,066 patients with mild to moderate IPF. Compared with placebo what was the reduction in annual rate of decline of FVC in both studies?

a. 125mL and 94mL
b. 167mL and 101mL
c. 199mL and 143mL
d. 243mL and 198mL
e. 278mL and 201mL

Answer: a. Treatment with nintedanib 150mg twice daily produced a statistically significant reduction in annual rate of decline of FVC in both studies (annual difference compared with placebo 125mL, p<0.001 and 94mL, p<0.001).

Question 5

Which one of the following should be performed before and during treatment with pirfenidone or nintedanib?

a. Full blood count
b. ECG
c. Blood pressure measurement
d. Renal function assessment
e. Hepatic function assessment

Answer: e. Elevated liver enzyme levels have been reported with pirfenidone and liver function tests should be carried out before and during treatment. Hepatic transaminase and bilirubin levels should be assessed before and periodically during treatment with nintedanib.