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Mr Mason
 Consultant Urologist
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 Level 6

PRR	FILE	
	TELL PT NORMAL	
CAK	TELL PT	Date: 29-JUN-2004
	TELL CON	: PR/MR /762159
PGM	30 JUN 2004	
VM		
NY	APPT	
	REPEAT	
NURSE	NOTES PLEASE	
	PRESCRIPTION	

Dear Mr Mason

Re: GRAHAM SHEPPARD
 6 BENEDICT CL TEIGNMOUTH DEVON TQ14 8FD
 NHS No: 6329131945

DoB: 11-DEC-1942

Patient Letter Copy: Not Copied - Third Party Involvement

Following our conversation about Mr Sheppard, I then reviewed him in the Haematology clinic later that day. As you will be aware he has myelofibrosis and is currently transfusion dependent requiring 2 to 3 units of blood every 4 weeks. He has previously been treated with a splenectomy, but as often happens in these patients, when the bone marrow is already fibrotic and the spleen is removed, he has developed massive hepatomegaly as a source of extra-medullary haemopoiesis. His disease has been in quite an accelerated phase with a high white count, occasional blasts in the peripheral blood and metabolic symptoms including weight loss. A previous search for possible bone marrow transplant failed to reveal any matched donors worldwide and he was also turned down by the Hammersmith Hospital for an autologous bone marrow transplant when their programme closed due to high transplant related mortality. He is currently on Thalidomide which I have just increased from 50 to 100 mg a day, as this has been shown to have some affect in controlling clonal haematological disorders. He initially had high doses of steroids as an adjunctive treatment but this has now reduced to 5 mg of Prednisolone a day. His long-term prognosis is poor and he is likely to either fully transform to acute leukaemia, succumb to bone marrow failure or develop a problem related to his transfusional iron overload. His ferritin is currently 3700 and he failed to tolerate Desferrioxamine infusions a year ago. Most recently I have discussed the oral chelating agent Deferiprone with him and our plan was to try this next month although it would take a considerable time before his ferritin came down to levels no longer considered at risk for end organ damage ie less than 1000.

In terms of his TURP, I made Mr Sheppard aware of a number of risk factors that he has for peri-operative problems. Firstly he is at increased risk of infection, having a primary marrow disorder and also having had a splenectomy in the past. Prophylactic antibiotics should therefore be considered. Secondly he is transfusion dependent and the operation would be best organised in the week following one of his top up transfusions. Thirdly and probably most importantly, people with myeloproliferative disorders are at risk both of thrombosis (arterial and

venous) and bleeding. Mr Sheppard's platelet count is currently normal, although his white count is quite high which will increase his whole blood viscosity. Perhaps most importantly however, PFA 100 analysis today shows that his platelets are quite dysfunctional and I think that his major peri-operative risk will be haemorrhage. I think that he needs transfusion of one adult therapeutic dose of platelets pre-operatively, and if he bled excessively despite this, we would have to consider using an injection of recombinant factor VIIa. All in all, I wonder if the Mount Stuart is the best place for his procedure just in case we did get into difficulties in controlling his haemostasis. Recombinant VIIa for example would only be available from the Blood Bank fridge in this hospital.

If, weighing up the risks and benefits, Mr Sheppard decides to go ahead with the procedure, perhaps you could let me know the timing so that we are prepared at our end. I hope this letter is helpful to you.

Yours sincerely



Dr Patrick Roberts
Consultant Haematologist

c.c. Dr C.A. Karakusevic - Bishopsteignton 13b

Ref: 762159/6329131945 /29-JUN-2004