

TEAM:

IUBMID:

28. Serum ferritin anytime prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

29. Specify (check only one):
1 <1000 ng/ml or µg/L
2 ≥1000 ng/ml or µg/L
8 Unknown

30. Was chelation therapy given at any time prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

31. Date started: Date unknown
Month Day Year

32. Predominant route (check only one):
1 IV
2 Subcutaneous
3 Intramuscular
7 Other, specify: _____
8 Unknown

33. Was Chelation therapy adequate (≥5 nights a week and ≥ hours a night)?
1 Yes 0 No 8 Unknown

34. Was a liver biopsy performed anytime prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

35. Date of most recent liver biopsy: Date unknown
Month Day Year

If Biopsy Report is available check here , attach copy and reference Q.34

36. Was hepatitis present?
1 Yes 0 No 8 Unknown

37. Specify:
1 Mild 2 Moderate 3 Severe 8 Unknown

38. Was siderosis present?
1 Yes 0 No 8 Unknown

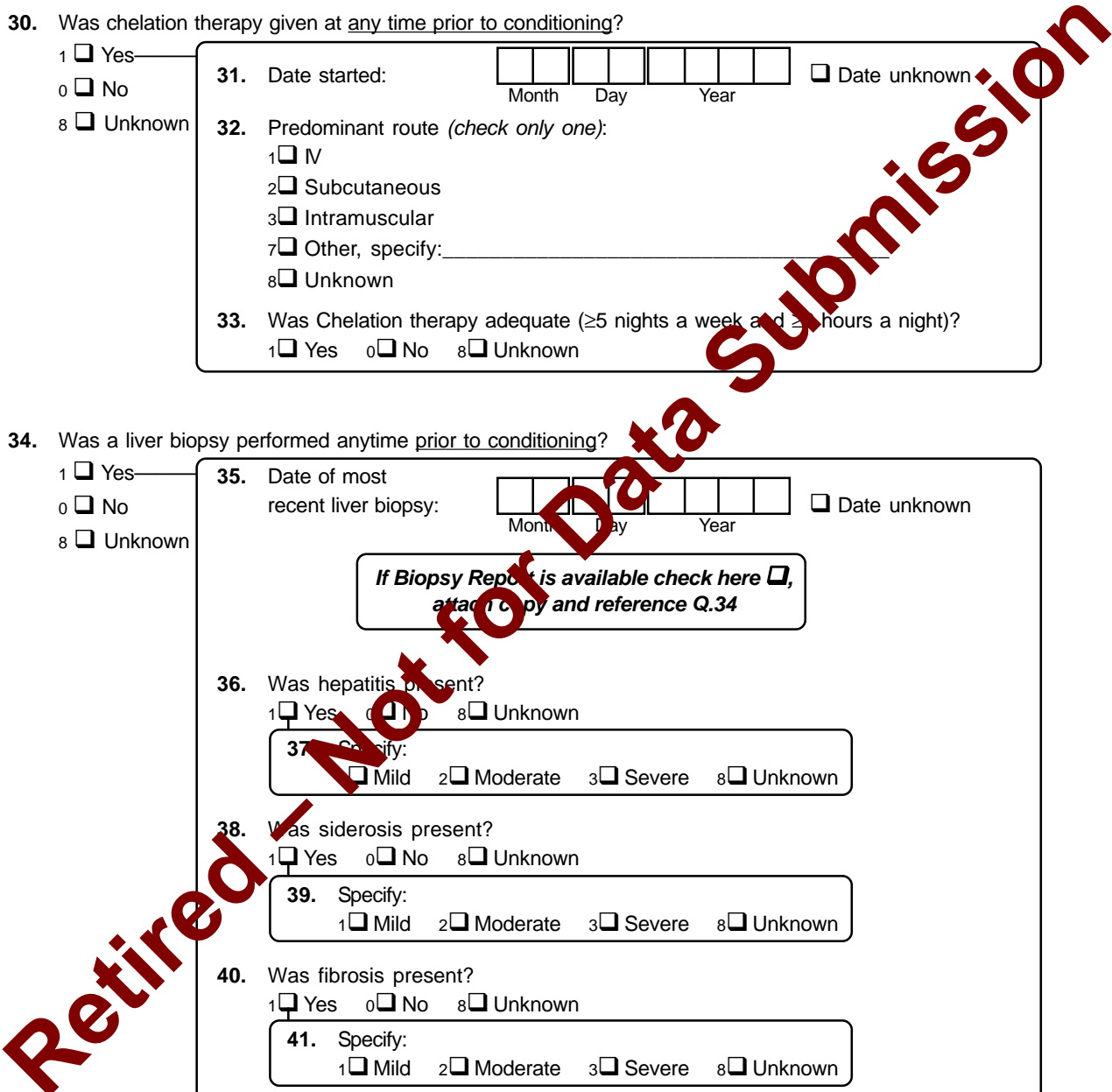
39. Specify:
1 Mild 2 Moderate 3 Severe 8 Unknown

40. Was fibrosis present?
1 Yes 0 No 8 Unknown

41. Specify:
1 Mild 2 Moderate 3 Severe 8 Unknown

42. Did serial liver biopsies show progressive disease?
1 Yes 0 No 7 N/A, serial biopsies not performed 8 Unknown

43. Hepatic iron content (HIC): mg/gm Unknown



TEAM:

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44. Were Pulmonary Function Tests (PFTs) performed anytime prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

If Report is available, check here , attach copy and reference Q.44

45. Specify results (check only one):

- 1 Normal
- 2 Stage 1 disease
- 3 Stage 2 disease
- 4 Stage 3 disease
- 5 Stage 4 disease
- 8 Unknown

Sickle Chronic Lung Disease Staging Criteria (Clinical)				
Markers	Stage 1	Stage 2	Stage 3	Stage 4
<i>Chest pain</i>	Recurrent substernal pain and chronic cough	Increased pain over Stage 1	Severe midline crushing chest pain	Severe and prolonged pain with dyspnea at rest
<i>Blood gases</i>	Normal oxygen saturation	Normal oxygen saturation	Hypoxemia with partial pressure of oxygen (70 mm Hg) during stable periods	Partial pressure oxygen (60 mm Hg) during stable periods
<i>X-ray</i>	Decreased distal pulmonary vascularity, hyperexpansion, evidence suggestive of increased interstitial markings	Diffuse, fine interstitial fibrosis	Pulmonary fibrosis	Severe pulmonary fibrosis
<i>Pulmonary function tests*</i>	Decreased FVC, TLC, FEV ₁ , and FEV ₁ /FVC ratio (mild, 80% of predicted normal, or 1 S.D. below normal)	Decreased FVC, FEV ₁ , TLC, and FEV ₁ /FVC ratio (moderate, 60% of predicted, or 2 S.D. below normal)	Decreased FVC, REV ₁ , TLC, DCO and FEV ₁ /FVC ratio (severe, 40% of predicted, or 3 S.D. below normal)	Patient frequently unable to complete testing due to degree of hypoxia
<i>ECG and ECHO</i>	Left ventricular preponderance persists	Balanced ventricular hypertrophy	Right ventricular hypertrophy and right atrial enlargement. Progressive increase in heart size	Severe right ventricular and right atrial hypertrophy. Ischemic T waves in V1 and V2, and p pulmonale
<i>Pulmonary artery pressure</i>	Normal	Normal	Borderline elevation or normal	Markedly elevated with pulmonary hypertension

Abbreviations: FVC = forced vital capacity, TLC = total lung capacity, REV₁ = forced expiratory flow rate

* These measurements are based upon common methods for comparison of reference values.

Retired - Not for Data Submission

TEAM:

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Disease symptoms anytime prior to conditioning:

Yes No Unknown

46. Vaso-occlusive pain requiring hospitalization within 2 years prior to transplant

47. Frequency: <3/yr ≥3/yr Unknown

48. Priapism

49. Number of episodes/yr:

50. Surgery? Yes No Unknown

51. Acute chest syndrome

52. Number of episodes in the 2 years prior to transplant:

53. Number of episodes in the patient's lifetime:

54. Requiring exchange transfusion? Yes No Unknown

Treatment for ACS:

Yes No Unknown

55. Transfusion of RBCs

56. Antibiotics

57. O₂

58. Intubation

59. Other, specify: _____

60. Prior stroke

61. More than one stroke? Yes No Unknown

62. Seizures

63. Osteonecrosis

Specify joints:

Yes No Unknown			Yes No Unknown				
64.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	67.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
65.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	68.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
66.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	69.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

70. Sickle neuropathy

Retired / Not for Data Submission

TEAM: [][][][]

IUBMID: [][][][][][][]

71. Did patient receive hydroxyurea at any time prior to transplant?

- 1 Yes
- 0 No
- 8 Unknown

If Report is available, check here , attach copy and reference Q.71

72. Start date: [][][][][][] Date unknown
Month Day Year

73. End date: [][][][][][] Date unknown

74. Was hemoglobin electrophoresis performed while on hydroxyurea?

- 1 Yes
- 0 No
- 8 Unknown

75. Date: [][][][][][] Date unknown
Month Day Year

Specify results:

76. Hb A1 [][] % Not tested while on hydroxyurea

77. Hb A2 [][] % Not tested while on hydroxyurea

78. Hb S [][] % Not tested while on hydroxyurea

79. Hb C [][] % Not tested while on hydroxyurea

80. Hb F [][] % Not tested while on hydroxyurea

81. Hb Other [][] % Not tested while on hydroxyurea
Specify: _____

82. Hb Other [][] % Not tested while on hydroxyurea
Specify: _____

If chronically transfused prior to transplant, provide pre-transfusion electrophoresis data

83. Did patient have gonadal dysfunction at any time prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

84. Was a brain MRI/MRA performed just prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

If MRI/MRA Report is available, check here , attach copy and reference Q.84

Cardiac Studies:

Yes No Unknown

85. 1 0 8 Pre-transplant EKG

Attach copy(ies), check here and reference Qs.85-86

86. 1 0 8 Pre-transplant Echocardiogram

87. Was a hemoglobin electrophoresis performed just prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

If Report is available, check here , attach copy and reference Q.87

88. Date: [][][][][][] Date unknown
Month Day Year

Specify results:

89. Hb A1 [][] %

90. Hb A2 [][] %

91. Hb S [][] %

92. Hb C [][] %

93. Hb F [][] %

94. Hb Other [][] %
Specify: _____

95. Hb Other [][] %
Specify: _____

If chronically transfused prior to transplant, provide pre-transfusion electrophoresis data

Retired - Not for Data Submission

TEAM:

IUBMID:

96. Primary reason for transplant (*check only one*):

- 1 Stroke
- 2 Acute chest syndrome
- 3 Recurrent vaso-occlusive pain
- 4 Recurrent priapism
- 5 Excessive transfusion requirements/iron overload
- 7 Other, specify: _____
- 8 Unknown

Retired – Not for Data Submission

TEAM:

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Posttransplant Information*

** To be completed 100 days posttransplant or at time of death if death occurred <100 days posttransplant, or immediately prior to start of high-dose therapy (conditioning) for second transplant if second transplant done <100 days after first transplant.*

97. Serum ferritin posttransplant:

- 1 <1000 ng/ml or µg/L
- 2 ≥1000 ng/ml or µg/L
- 8 Unknown

98. Was chelation therapy given posttransplant?

- 1 Yes
- 0 No
- 8 Unknown

99. Is patient still receiving chelation therapy or undergoing phlebotomy?

- 1 Yes
- 0 No
- 8 Unknown

100. Date ended: Date unknown
Month Day Year

Disease symptoms posttransplant:

Yes No Unknown

101. 1 0 8 Vaso-occlusive pain requiring hospitalization

102. Frequency: 1 <3/yr 2 ≥3/yr 8 Unknown

103. 1 0 8 Priapism

104. Number of episodes/yr:

105. Surgery posttransplant? 1 Yes 0 No 8 Unknown

106. 1 0 8 Acute chest syndrome

107. Number of episodes posttransplant:

108. Requiring exchange transfusion? 1 Yes 0 No 8 Unknown

Treatment for ACS:

Yes No Unknown

109. 1 0 8 Transfusion of RBCS

110. 1 0 8 Antibiotics

111. 1 0 8 O₂

112. 1 0 8 Intubation

113. 1 0 8 Other, specify: _____

114. 1 0 8 Stroke

115. More than one stroke? 1 Yes 0 No 8 Unknown

116. 1 0 8 Seizures

117. 1 0 8 Osteonecrosis

Specify joints:

Yes No Unknown

Yes No Unknown

118. 1 0 8 Ankle 121. 1 0 8 Shoulder

119. 1 0 8 Hip 122. 1 0 8 Spine

120. 1 0 8 Knee 123. 1 0 8 Other, specify: _____

124. 1 0 8 Sickle nephropathy

TEAM:

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125. Was a brain MRI/MRA performed posttransplant?

- 1 Yes
- 0 No
- 8 Unknown

If MRI/MRA Report is available, check here , attach copy and reference Q.125

Cardiac Studies:

Yes No Unknown

126. 1 0 8 EKG posttransplant

Attach copy(ies), check here and reference Qs.126-127

127. 1 0 8 Echocardiogram posttransplant

128. Was a hemoglobin electrophoresis performed posttransplant?

- 1 Yes
- 0 No
- 8 Unknown

If Report is available, check here , attach copy and reference Q.128

129. Date: Date Unknown
Month Day Year

Specify results:
(include all posttransplant electrophoresis results with dates performed)

- 130. Hb A1 %
- 131. Hb A2 %
- 132. Hb S %
- 133. Hb C %
- 134. Hb F %
- 135. Hb Other %

Specify: _____

If more than one posttransplant electrophoresis performed, copy this page.

136. Did patient have gonadal dysfunction posttransplant?

- 1 Yes
- 0 No
- 8 Unknown

137. Patient's current disease status at date of last contact (see pg 1 of 002-CORE)
(check only one)

- 1 Cured: no electrophoresis (Hb S \leq 50%) and clinical symptoms (see **Qs.101-124**) absent
- 2 Disease recurred: Hb S >50% and absence of clinical symptoms (see **Qs.101-124**)
- 3 Disease recurred: Hb S >50% and presence of clinical symptoms (see **Qs.101-124**)
- 8 Unknown

