TAMOF children (Plasma Exchange=60 vs. Standard Therapy=21) PELOD Score vs Days on Study: Max PELOD used at all time points after death. PELOD Score vs Days on Study 40 40 Plasma Exchange
Standard Therapy Plasma Exchange
Standard Therapy 35 35 30 30 PELOD Score 25 PELOD Score 25 20 20 15 15 10 10 5 5 0 0 Day: 3 5 Day 6 (47)(#Plasma Exchange) (60) (52)(48)(46)(45)(39)(37)(#Plasma Exchange) (60) (55)(55)(55)(54)(49)(#Standard Therapy) (21) (13)(13)(13)(#Standard Therapy) (21) (15)(15)(14)(14)(14)(16)(16)(16)(15)Days on Study Days on Study Platelet Count vs Days on Study ADAMTS-13 Activity vs Days on Study (Plasma Exchange=43 vs. Standard Therapy=13) 180 Plasma Exchange
Standard Therapy 100 Plasma Exchange
Standard Therapy 160 90 Platelet Counts(*1000 cells/uL) 140 80 ADAMTS-13 Activity (%) 120 70 100 60 50 80 40 60 30 40 20 20 10 0 Day: Day 28 (#Plasma Exchange) (58) (56)(50)(49)(43)(40)(29)(34)(32)(30)(14)(51)(#Plasma Exchange) (22)(#Standard Therapy) (21) (#Standard Therapy) (13)(7) (19)(19)(18)(18)(16)(17)(11)(12)(4) Days on Study Days on Study vWF Antigen vs Days on Study vWF Ristocetin Cofactor Activity vs Days on Study (Plasma Exchange=43 vs. Standard Therapy=13) (Plasma Exchange=43 vs. Standard Therapy=13) 280 360 Plasma Exchange 340 260 vWF Ristocetin Cofactor Activity (%) Standard Therapy 320 240 300 220 280 260 200 $\frac{240}{240}$ 180 220 160 200 140 180 160 120 140 100 120 80 100

80

60

40

20

15

(22)

28

(13)

Day

(#Plasma Exchange)

(#Standard Therapy)

(29)

(30)

(29)

(12)

Days on Study

(29)

15

(23)

28

(14)

(4)

60

40

20

Day:

(#Plasma Exchange)

(#Standard Therapy)

Plasma Exchange

standard Therapy

(29)

(35)

(11)

(33)

Days on Study

(30)