

**Unpublished PPB Registry Compilation of PPB Patients Treated with High-Dose Chemotherapy and Stem Cell Reconstitution**

<b>ASCR Reference</b> and Site of care	<b>Description of Case</b>	<b>Outcome</b> (post ASCR)
Schmaltz: Med Pediatr Oncol 1995; 25:479-484 (Freiberg and Essen, Germany)	PPB, prob III. Partial resection. 1st rx = CWS 91, German Soft Tis Sarc Rx, w improve. After 3 mo rx, lung progression. Another partial resect; then carbo & vp @ 450/m2 each. Then HDCT + ASCR: melphalan (180 mg/m2) etoposide (40 mg/kg) carboplatin (1500 mg/m2). CT = regression. Then add idarubicine (24 mg/m2). Then lung recurrence ~1-2 mo later & DOD in 6 wks more.	DOD
de Castro J Pediatr Hematol/Oncol 2003; 25:78-81 (Porto Alegre, Brazil)  Registry Case	PPB, Type III. Bx. VAC(V = 1.5 mg/m2; A = 1.5mg/m2; C = 2.2gm/m2) X 3 = 25% decr. Change to ICE (I = 3000mg/m2 D1-3; C = 400mg/m2 D1&2; E = 166 mg/m2 D1-3) X 4 = 75% PR. Surg = partial resect. Then ICE X 2. Then HDCT + ASCR: Melphalan(140mg/m2 D"-5, 70 D"-4"); etop(160mg/m2 D"-6" to "-3"; carbo(300mg/m2 D"-6" to "-3"). 3 mo post ASCR = NED. 1 mo later recurrence. DOD 20 mo fr dx and 9 mo fr ASCR.	DOD
Bekassy: Eur Paediatr Haematol Oncol 1984; 1:147-155 (Lund, Sweden) (Bekassy ltr to ed Med Pediatr Oncol 1997; 28:75-76 does not add any pt)	1982 case. Prob PPB. Lung path after chemo. Skull met at dx and presenting sx! First dx = bone Ewings + lung mets. 1st rx = Rosen T-11. Some response. Then lung resection. Then more bone mets. Then HDCT + autol BM: HD melphalan. Early lung recurrence and DOD 9 mo p dx.	DOD
Bongo: Bull Cancer 1996; 83:877-881 (Marseille, France)	PPB, prob III. Surg = partial resect. 1st rx = SIOP MMT 89, group E:CE then IVA then IVP16 [doses: carbo(500mg/m2), epibubicin(150mg/m2), Vcr(1.5mg/m2) then ifos(3gm/m2), actino(1.5mg/m2), Vcr(same) then ifos(same), etop(300mg/m2), Vcr(same)]. 3 courses of 3 sequences in 6 mo: small resid disease. 2nd surg = microscopic resid. HDCT + ASCR: melphalan(180mg/m2), etop(200mg/m2 X 5), carbo(350mg/m2 X 5). Status NED 7 yr post ASCR. (f/u status per de Castro paper)	NED 7 yr
Kusafuka: Pediatr Hematol Oncol 2002; 19(2):117-28. (Osaka, Japan)	PPB, prob III. No initial bx. Chemo = cisplat(90mg/m2), etoposide(500mg/m2), pirarubicin(40mg/m2), cyclophos(1200mg/m2) X 1. Effect = "partial, but inadequate response". Surg = subtotal resect. Add'l unspecified chemo. Then HDCT + ASCR X 2: HDCT#1 = ifos(12,500mg/m2), melphalan(210mg/m2). 109 d later: HDCT#2 = busulphan(20mg/kg), thiotepa(800mg/m2). Status: NED 3 yr post 2nd ASCR.	NED 3 yr
Registry Case (USA)	PPB II. Surg = presumed total resection. 1st chemo = VAC alt with cisplat/doxo X 9: doses: Vcr(2.0mg/m2 wkly x 3), actino(1.25mg/m2), cpm(1.2gm/m2), doxo(75mg/m2), cisplat(20mg/m2 x 5d). Primary lung recur @ 22 mo. Surg = lobectomy. 2nd chemo = ifos/etoposide alt with topotecan/cpm x 4 mo. Then XRT = 1200cGy whole lung + tumor bed boost total 4100. Then HDCT + ASCR: HD cpm, busulfan (30 mg/m2 X 16 in 4 days) and melphalan(180 mg/m2). Status NED 53 mo post dx, 24 mo post ASCR.	NED 49 mo as of 4/05
Registry Case (USA)  (CNS Recurrence)	PPB III. Gross total resect. 1st chemo = doxo, cyclophos X 2. CNS recur @ 60 months. Surgery. Then HDCT + ASCR + CNS XRT. HDCT = HD cyclophos X 2. XRT: 3000cGy whole brain & 1500 tumor boost. Status DOD 12 mo post ASCR.	DOD 12 mo after ASCR

Registry Case (USA)	PPB, probable Type II. Resection with large recurrence at 2 mo; chemo X 5 mo: V, AMD, carboplat, epirubicin + protectant, and etoposide(doses unk). HD CPM for harvest. Chemo X 2-3 mo:V, AMD, Ifos, Adria, cyclophos (doses unk).XRT: 1500cGy to hemithorax. HDCT = HD etoposide carbo, and esc doses of cyclophos + ASCR	NED 21 mo
Registry Case (USA)	PPB II. Gross total resect. 1st chemo =VAC/V x 1 course, then VAC x 13 courses. Local recurrence @ 35 mo. Presumed treated with Etop/Ifos alt. with Cisplat/Doxo, then ASCR. ASCR completed 8 mo after relapse. Status NED 50 days post ASCR.	recur 11/2003 ASCR 7/2004 NED 3/11/05
Registry Case (USA) <b>(CNS Recurrence)</b>	PPB III. Gross total resect 3 days after biopsy.1st chemo = VAC alt. with Cisplat & Doxo. CNS recur at 13 mo. <b>Treatment details and current status pending.</b> <u>Rx after CNS resection and before ASCR:</u> ICE x 1, then oral VP x 2 weeks. <u>Conformal XRT</u> to frontal lobes: 3960 cGy, 22 fx, over 30 days elapsed <u>ASCR RX:</u> Carboplat: AUC target 7 per day ( <b>dose pending</b> ) days -8, -7, -6 Thiotepa: 200 mg (10 mg/kg) IV days -5, -4, -3 VP16: 170 mg (8.3 mg/kg) IV days -5, -4, -3 Infusion: day 0.	recur 12/2003 NED pr MD 4/15/05
German CWS Study Kirsch et al; SMS 2005 Congress	Case #5: "EVAIA" rx non-responder, followed by high-dose stem cell transplant (details unknown to the Registry)	DOD 9 mo p PPB dx