International Pleuropulmonary Blastoma (PPB) Treatment and Biology Registry

Types I, II and III PPB

APPENDIX XII: REPORTING SCHEDULE AND CASE REPORT FORMS
## APPENDIX XII: REPORTING SCHEDULE AND CASE REPORT FORMS

### IPPBTR-1 TYPES I, II & III DATA AND SPECIMEN SUBMISSION SCHEDULE:

<table>
<thead>
<tr>
<th>Required Materials</th>
<th>Send to:</th>
<th>At time of Enrollment</th>
<th>End of RT</th>
<th>End of each phase</th>
<th>At relapse/progression/secondary malignancy</th>
<th>At time of each surgery</th>
<th>During follow-up</th>
<th>At death</th>
</tr>
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<tbody>
<tr>
<td>Enrollment Form</td>
<td>IPPBTRBRO *</td>
<td>X</td>
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<tr>
<td>Representative Formalin-Fixed Paraffin Blocks of Tumor Material (A)</td>
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<td>X (A)</td>
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<td>Pathology Report</td>
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<td>Surgery Reporting Form</td>
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<td>End of Phase/ Course Report and Completed Roadmap*</td>
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<td>Imaging Reports and Electronic Digital Images</td>
<td>IPPBTRBRO</td>
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<td>X</td>
<td>X</td>
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<td>RT Course Form AND RT Treatment Summary*</td>
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<td>X (C)</td>
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* INTERNATIONAL PLEUROPULMONARY BLASTOMA TREATMENT AND BIOLOGY REGISTRY OFFICE (MINNEAPOLIS, MN)

**REQUIRED IF SECONDARY MALIGNANCY IS SPECIFICALLY AML/MDS

* REQUIRED ONLY FOR PATIENTS TREATED PER REGISTRY TREATMENT RECOMMENDATIONS

(A) If blocks absolutely cannot be sent, then send: (a) 1 H&E section of all available blocks, and (b) 10 unstained sections (on plus-charged, polarized slides) for immunoperoxidase studies from 2 representative blocks, and (3) 2 H&E slides from the same blocks. See protocol Appendix V

(B) Within 1 year of the end of Treatment and Biology Registry VAC (for T-I) or IVADO (for T-II & III) regimen guideline therapy, then within 1 year of completion of last form.

(C) To cover period from last follow-up form submitted to date of death.
## IPPBTBR-1: Eligibility Worksheet

<table>
<thead>
<tr>
<th>Institution:</th>
<th>Patient Name:</th>
</tr>
</thead>
</table>

| Informed consent signed? | 1 = Yes  
2 = No, Ineligible |
|--------------------------|-------------------|

<table>
<thead>
<tr>
<th>Date consent (and assent, if required) signed?</th>
<th>/</th>
<th>/</th>
<th>/</th>
</tr>
</thead>
<tbody>
<tr>
<td>month</td>
<td>day</td>
<td>year</td>
<td></td>
</tr>
</tbody>
</table>

| Patient is less than 21 years of age? | 1 = Yes  
2 = No, Ineligible |
|--------------------------------------|-------------------|

| Patient has newly-diagnosed Type I, II or III pleuropulmonary blastoma (PPB)? OR | 1 = Yes  
2 = No, Ineligible |
|-------------------------------------------------------------------------------|-------------------|

| Patient has recurrent PPB? | 1 = Yes  
2 = No |
|---------------------------|---------|

| Female patients only: Patient is of childbearing age and pregnancy test is negative? | 1 = Yes  
2 = No, Ineligible  
3 = N/A. Patient is male, or is female but not of childbearing age |
|-----------------------------------------------------------------------------|-------------------|

| Has patient been informed of the PPB Genetic study? | 1 = Yes  
2 = No (if not, please refer them to the study website,  
http://www.ppbgeneticstudy.org/) |
|--------------------------------------------------|-------------------|

Fax this completed form to the International PPB Treatment and Biology Registry office, 612-813-7108.  
For questions, please contact the office at 612-813-7115.
Patient Questions:
Are there any other major illnesses in the patient?:

Has this child ever had a chest x-ray or chest CT prior to the current situation?:

Has this child ever had a pneumothorax?:

Has this child had any pneumothorax?:

Has this child had any lung cysts diagnosed prior to PPB?:

Has this child had any kidney cysts?:

Has this child had any kidney cysts?:

Has this child had any thyroid nodules/surgery?:

Does this child have birth defects/syndromes/dysplasias (such as VACTERL, Fragile X, etc, etc)?:

If you answered YES to any of the above questions, please provide details below:

Family Questions: Has any brother, sister, parent, relative had any of the following:

Pneumothorax?:

Kidney cysts?:

Ovarian Tumors; testicular tumors?:

Lung cysts?:

Thyroid nodules or tumors?:

Childhood Cancers?:

If you answered YES to any of the above questions, please provide details below:

Has this child’s mother or father had previous marriages?

Are there any “half-siblings” of the patient?:

Are there any of the above findings in any half-siblings?: If YES, please provide details above.

*The questions in this survey emphasize medical conditions found in the PPB Family Tumor Susceptibility Syndrome
IPPBTBR-1 TYPES II & III TREATMENT END-OF-PHASE/COURSE REPORT FORM

Phase #: ___   Course: ___

Phase 1, Course 1 = (weeks 0-2)
Phase 1, Course 2 = (weeks 3-12)
Phase 2, Course 1 = (weeks 12-24)
Phase 3, Course 1 = (weeks 24-42)

Number of days hospitalized: ______   Number of hospital admissions: ______

Height: ______ cm   Weight: ______ . ___ kg   BSA: ___ . ___ m²

Date course began: ______ / ______ / ______
month   day   year

Date course ended: ______ / ______ / ______
month   day   year

Total Dose for this Course:

Ifosfamide: ______ . ___ ___ mg
Vincristine: ______ . ___ ___ mg
Actinomycin D: ______ . ___ ___ mg
Cyclophosphamide: ______ ______ ___ . ___ ___ mg
Doxorubicin: ______ . ___ ___ mg
Mesna: ______ . ___ ___ mg

Did the patient have surgery during this course? (1= Yes; 2= No): ___
If yes, Please complete the IPPBPS Surgery Reporting Form.
Phase #:   Course:  

End-of-Course Overall Objective Response Prior to Surgery: For all patients prior to surgery. (Leave blank if no surgery was performed. Response is always recorded in comparison to the tumor size at diagnosis, not at the last evaluation.)

Response:   
1 = Complete response/remission (CR): Disease free, all sites  
2 = Partial response/remission (PR): 50% decrease in the sum of the products of the maximum perpendicular diameters of all measurable lesions for 4 weeks, no new lesions  
3 = No response/remission (NR) or stable disease (SD): <50% decrease, and no new lesions  
4 = Progressive disease (PD): 25% increase and/or the appearance of new lesions, OR Relapse/recurrence (R): Appearance of new lesions or reappearance of old lesions for patients in CR.  
0 = No evaluation performed

Date of evaluation:   /   /   

month     day     year

End-of-Course Overall Objective Response: For all patients (MUST BE COMPLETED). ** (Please record overall clinical response even if imaging was not performed. Response is always recorded in comparison to the tumor size at diagnosis, not at the last evaluation.)

Response:   
1 = Complete response/remission (CR): Disease free, all sites  
2 = Partial response/remission (PR): 50% decrease in the sum of the products of the maximum perpendicular diameters of all measurable lesions for 4 weeks, no new lesions  
3 = No response/remission (NR) or stable disease (SD): <50% decrease, and no new lesions  
4 = Progressive disease (PD): 25% increase and/or the appearance of new lesions, OR Relapse/recurrence (R): Appearance of new lesions or reappearance of old lesions for patients in CR.  
0 = No evaluation performed

**NOTE: For patients who have had surgery this is the response following surgery.

Date of evaluation:   /   /   

month     day     year

Therapy Administration:

Was therapy administered as per the Treatment and Biology Registry IVADo regimen guideline, including specified modifications for toxicity?  
(1= Yes; 2= No):   

(NOTE: Patients terminating the Treatment and Biology Registry IVADo regimen guideline for relapse/progression, or death, are considered to have followed Registry IVADo regimen guideline?)

If no, What was the nature of the difference from the Treatment and Biology Registry IVADo regimen guideline?:   
1= Deviation from the Treatment and Biology Registry IVADo regimen guideline, but Registry-suggested therapy will generally be followed henceforth  
2= Abandonment of the Treatment and Biology Registry IVADo regimen guideline therapy-no further attempt to follow Registry-suggested therapy  
3= N/A, patient has completed the Treatment and Biology Registry IVADo regimen per guidelines.
IPPBTBR-1   TYPES II & III TREATMENT
END-OF-PHASE/COURSE REPORT FORM

Phase #: __    Course: __

Off Therapy:

Was the patient taken off the Treatment and Biology Registry IVADo regimen guideline this course (including patients completing Registry-suggested therapy)?: (1= Yes; 2= No): ___

If yes, reason off the Treatment and Biology Registry IVADo regimen guideline this course: ___

00 = No evaluation performed.
01 = Excessive toxicity, physician discretion
02 = Other physician discretion
03 = Best interest of patient/other than toxicity, physician discretion
04 = Parent/patient discretion
05 = Progression/No response per IPPBTBR-1 regimen
06 = Completed Registry-suggested therapy
07 = Review histology made patient ineligible
08 = Unable to contact patient (lost)
09 = Other, Specify: ________________________________
10 = Death

Form completed by:
Name: _________________________  : ___________________________ |__|__| / |__|__|__|__|
(Please print) (Signature) month day year

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SURGICAL REPORTING FORM

To be submitted for any surgery AFTER beginning treatment.

Time Surgery performed:  
1 = During Phase 1, Course 1 (Types II and III only)  
2 = During Phase 1, Course 2 (Types II and III only)  
3 = During Phase 2, Course 1(Types II and III only)  
4 = During Phase 3, Course 1(Types II and III only)  
5 = Other

Date of Surgery:__ /__ /__
month day year

Pre-operative Status of primary site (clinical):  
1= Complete Response (CR)  
2= Partial Response (PR)  
3= No Response (NR)  
4= Increasing Disease (INC)  
9= Unknown (UNK)

Surgical Procedure:  
1 = Lobectomy  
2 = Cystectomy  
3 = Wedge/segment  
4 = Pneumonectomy  
5 = Tumorectomy  
6 = Biopsy*  
7 = Other, non chest site. Specify:____________________________________

Operative Site:  
1 = Visceral Pleura  
2 = Parietal Pleura  
3 = Chest Wall/Rib Cage  
4 = Diaphragm  
5 = Mediastinum  
6 = Pericardium  
7 = Major Vesses Surrounded  
8 = Other, non chest site. Specify:____________________________________

Degree of Resection:  
1 = Biopsy*  
2 = Subtotal  
3 = En Bloc  
4 = Most/known residual  
5 = Gross Total  
6 = Piecemeal  
7 = Other. Specify:____________________________________

*If Surgery to Primary Was Biopsy only:

Type of biopsy -  
Open or closed:  
1= Open; 2 = Closed; 9 = N/A  
Needle or Incisional:  
1= Needle; 2 = Incisional; 9 = N/A  
Fine needle biopsy:  
1= Yes; 2 = No; 9 = N/A

Reason for Biopsy are (check all that apply):

[ ] Clinically Unresectable  [ ] Distant metastases  
[ ] Initial Rx. Plan  [ ] Other, specify: ____________________________
Known Spill/Rupture? □ 1 = Yes; 2 = No
Nodes Involved? □ 1 = Yes; 2 = No
Empyema? □ 1 = Yes; 2 = No
Was this Surgery Before Neo-Adjuvant Chemotherapy? □ 1= Yes; 2= No
Was there Vascular Invasion? □ 1 = Yes; 2 = No
What is the status of the patient now that this surgical procedure has been completed? □
1= Complete Response (CR)
2= Partial Response (PR)
3= No Response (NR)
4= Increasing Disease (INC)
9= Unknown (UNK)

List Other Findings or Procedures of Importance, i.e., Intestinal Resection, Application of Interstitial Irradiation, etc.:
_____________________________________________________________________________________
_____________________________________________________________________________

Form completed by:
Name: ____________________________ : ____________________________ |__|__| / |__|__| / |__|__|__|__|
(Please print) (Signature) month day year

Fax this completed form, along with your institution’s pathology report, to the International PPB Treatment and Biology Registry office, 612-813-7108.
For questions, please contact the office at 612-813-7115
TUMOR BIOLOGY CHECKLIST

Were preserved pathology materials sent to the IPPBTBR for Tumor Central Review?

___ 1= Yes; 2= No

If no, Reason not sent: ___

1= Biopsy/surgery at outside hospital
2= No excess tissue available for IRS biology studies
3= RMS not suspected at time of surgery
4= Pathology unaware of protocol requirement
5= Patient/parent refusal
6= Oversight
7= Reason not reported
9= Other,
Specify:___________________________________

Was frozen tissue and other Biologic Materials sent to the IPPBTBR for Biologic Studies?

___ 1= Yes; 2= No

If no, Reason not sent: ___

1= Biopsy/surgery at outside hospital
2= No excess tissue available for IRS biology studies
3= RMS not suspected at time of surgery
4= Pathology unaware of protocol requirement
5= Patient/parent refusal
6= Oversight
7= Reason not reported
9= Other,
Specify:___________________________________

Form completed by:

Name: ___________________________  ___________________________
(Please print)  (Signature)  month  day  year

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RADIATION COURSE FORM

Start Date of Radiation Therapy: |__|__| / |__|__| / |__|__|__|__|

End Date of Radiation Therapy: |__|__| / |__|__| / |__|__|__|__|

Description of Radiation Course: |__| 1= Primary chest disease
2= Recurrence/Metastatic chest disease
3= Brain Metastasis
4= Non-Chest/Non-Brain metastatic disease
5= Palliative

Radiation Modality: |__| 1= External beam
2= Conformal External beam
3= Intracavitary radioactive liquid
4= Gamma knife (stereotactic radiosurgery)
5= Radioactive Seeds
6= Other. Specify: _______________________________________________________

Course description:
Preferred course description = total cGy, # of fractions, field description, elapsed days [= “duration”] (e.g.: 400 cGy in 20 fx to whole brain in 26 days).

Site Location for Radiation Therapy: ____________________________________________

Primary Treating Radiation Oncologist: ____________________________________________

Form completed by:

Name: ___________________________ : _____________________________ |__|__| / |__|__| / |__|__|__|__|

(Please print) (Signature) month day year

Fax this completed form, along with the Radiation Therapy Summary Report, to the International PPB Treatment and Biology Registry office, 612-813-7108.

For questions, please contact the office at 612-813-7115.
FOLLOW UP FORM

Time period covered by this form:

From:  |__|__| / |__|__| / |__|__|__|__|
   month       day            year

To:  |__|__| / |__|__| / |__|__|__|__|
   month       day            year

Current Known Status:  |__|  1= No Evidence of Disease
                      2= Dead of Disease
                      3= Alive with Disease
                      4= In Primary Treatment
                      5= In Recurrence/Metastatic Treatment
                      6= Lost to Follow-up

□   Did this patient relapse/progress during this reporting period?
   1= Yes; 2= No   If yes, Complete Relapse/Progression/Second Malignant Neoplasm Form

□   Was this patient diagnosed with a second malignancy during this reporting period?
   1= Yes; 2= No   If yes, Complete Relapse/Progression/Second Malignant Neoplasm Form

□   Did the patient die during this reporting period?
   1= Yes; 2= No   If yes, Complete Relapse/Progression/Second Malignant Neoplasm Form

□   Is this patient now lost to follow-up?
   1= Yes; 2= No
   If yes, complete the following:

□□□□ Number of attempts to contact patient

Methods tried, check all that apply:
   □   Phone
   □   Letter
   □   Family doctor
   □   Other, Specify: _______________________________________

Form completed by:
Name: __________________________ : ______________________________ |__|__| / |__|__| / |__|__|__|__|
   (Please print)     (Signature) month   day   year

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IPPBTBR-1  Treatment = IVADo

RELAPSE/PROGRESSION/SECOND MALIGNANT NEOPLASM FORM

Date of Relapse/Progression/Second Malignant Neoplasm:  [month] / [day] / [year]

How was Relapse/Progression/Second Malignant Neoplasm Diagnosed? Check all that apply:

☐ Simple X-Ray ☐ Biopsy
☐ XR-CT ☐ Surgery
☐ MRI ☐ Physical Exam
☐ XR – Nuc. Scan ☐ Other. Explain ______________________________

If Relapse/Progression: 1= Yes; 2= No

Local recurrence:  ☐
Regional nodes:  ☐
Metastases:  ☐
If yes, New?:  ☐

Surgically proven:  ☐ (If yes, Send OP/Path or BM Reports, and complete the IPPBPS Surgery Reporting Form)

Sites: ______________________________________________________________________

If Second Malignant Neoplasm:

Type: ______________________________________________________________________

Send OP/Path or BM Reports and AML/MDS Report if applicable.

Sites: ______________________________________________________________________

Notes:

- If patient has a Relapse/Progression on the SAME date as a Second Malignant Neoplasm use one form for all data.

- If patient has a Relapse/Progression on one date and a Second Malignant Neoplasm on another date complete two forms -- one for the Relapse/Progression and another for the Second Malignant Neoplasm.

Form completed by:

Name: ___________________________ : _____________________________ [month] / [day] / [year]

(Please print) (Signature)
DEATH REGISTRATION FORM

Date of Death: _____ / _____ / ______

Was autopsy done? 1 = Yes; 2 = No; 9 = Unknown

CAUSE(S) OF DEATH:

*Was this factor a significant* contributory factor in death?
(*A factor is "significant" if it played an integral part in the patient’s death)

Codes: 1 = Yes; 2 = No; 9 = Unknown

Give details:

1. Progressive disease
   Site: _________________________________
   Other: _________________________________

2. Infection
   Organism: _________________________________
   Other: _________________________________

3. Hemorrhage
   Site: _________________________________
   Other: _________________________________

4. Toxicity
   Site: _________________________________
   Other: _________________________________

5. GVHD
   Site: _________________________________
   Other: _________________________________

6. Operative complications
   _________________________________

7. Unrelated to original
diagnosis & treatment
   (e.g.; drowning)
   _________________________________

8. Other
   _________________________________

Did the patient die within 3 months of the end of IPPBPS protocol therapy or while on IPPBPS protocol therapy? (1 = Yes; 2 = No)

If yes, how were the side effects of protocol therapy related to the patient's death?

1 = Not related
2 = Minor contribution to death
3 = Major contribution to death
4 = Exclusive cause of death

**If 2, 3 or 4, Explain the relationship:

If you had to pick one, which of the above (1-8) was the main reason the patient died?

Form completed by:

Name: ____________________________
(Please print)

Signature: ____________________________

Institution Name: ____________________________

Registry ID Number: ____________________________

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Date Cyst(s) First Noted: ___/___/____

When cysts were first noted, did the patient have a pneumothorax? 1= Yes; 2= No; 9= Unknown

Number of Cysts: ___

Location of lung cyst(s): Check all that apply:

- □ Upper
- □ Mid
- □ Lower
- □ Pleura
- □ Lingula
- □ Lingula
- □ Diaphragm
- □ Pleura
- □ Unspecified
- □ Right
- □ Left
- □ Unspecified

Size of Cysts:

|  |  | cm |  | cm |  | cm |  | cm | Cyst Location: ____________________
|---|---|----|---|----|---|----|---|----|
|  |  | cm |  | cm |  | cm |  | cm | Cyst Location: ____________________
|  |  | cm |  | cm |  | cm |  | cm | Cyst Location: ____________________
|  |  | cm |  | cm |  | cm |  | cm | Cyst Location: ____________________
|  |  | cm |  | cm |  | cm |  | cm | Cyst Location: ____________________
|  |  | cm |  | cm |  | cm |  | cm | Cyst Location: ____________________
|  |  | cm |  | cm |  | cm |  | cm | Cyst Location: ____________________

Cyst Description: 1= unilocular; 2= multilocular; 9= Unknown

Method of Measurement: ___

1= MRI  2= CT Scan  3= X-ray  4= Ultrasound  5= Radionuclide Scan  6= Physical Exam  9= Other (specify: ________________)

Form completed by:

Name: __________________________: __________________________ |
(Please print) (Signature) month day year

IPPBTBR-1  Page 1 of 1  Sept 2009
IPPBTBR-1  Treatment = IVADo

DIAGNOSTIC SPECIMEN TRANSMITTAL FORM

See protocol sections Appendix V.

Date of Surgery: __ / __ / __ 

Surgical Pathology #: __________

Referring Pathologist

Name of Referring Institution

Referring Institution Address

City  State  Zip/Postal Code

Country

Telephone #  Fax #

Contact Person’s Email

Include the following with your submission:

• A complete set of hematoxylin and eosin-stained slides
• A complete pathology report
• One representative paraffin block or 15 unstained sections
• Your contact information including phone number, fax, and e-mail address
• This form, filled out in its entirety

Comments/Instructions: __________________________________________________________

Shipping

Arrangements should be made for Federal Express pick-up according to the usual institutional procedure. Call ahead (612-813-7115) to receive a Federal Express account number for your shipment.

Specimens should be shipped to:

International Pleuropulmonary Blastoma Registry
Children’s Hospitals and Clinics of Minnesota
2545 Chicago Ave. S.
Suite 412
Minneapolis, MN 55404 USA
Telephone: 612-813-7115
Email: gretchen.williams@childrensmn.org

FOR INTERNATIONAL SHIPMENTS:

Please label outside of the package with the following: “The package contains preserved, non-infectious human tissue on glass slides or in paraffin wax for medical diagnosis purposes.”