**Supplemental Material**

**Validity of ESC risk assessment in idiopathic pulmonary arterial hypertension in China**

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**Supplementary Figure S1.** Kaplan-Meier survival estimates of all patients with IPAH at baseline.



**Supplementary Figure S2.** Patient selection flow chart.

BNP: brain natriuretic peptide; IPAH: idiopathic pulmonary hypertension; 6MWD: 6-minute walking distance; NT-proBNP: N-terminal fragmental of pro-brain natriuretic peptide; RA: right atrium; VE/VCO2: ventilatory equivalents for carbon dioxide; VO2: oxygen consumption; WHO FC: World Health Organization functional class.



**Supplementary Figure S3.** Forest plot based on the prognostic values of 6-min walking distance (6MWD), World Health Organization (WHO) function class (FC), brain natriuretic peptide (BNP) or N-terminal fragment of proBNP (NT-proBNP), right arterial pressure (RAP), cardiac index (CI) and mixed venous oxygen saturation (SVO2) in the Intermediate risk (IR) and High risk (HR) groups. Values for the variables were obtained from re-evaluation. The reference value is from the respective Low risk group.

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| **Supplementary Table S1. PAH-targeted treatment initiated within 3 months after PAH diagnosis and re-evaluation** |
| **Treatment** | **Baseline** **(392)** | **Re-evaluation (286)** |
| **Monotherapy, n (%)** |  |  |
| ERA\* | 56 (14) | 38 (13) |
| PDE5-I# | 186 (48) | 107 (37) |
| Prostanoid¶ | 16 (4) | 10 (3) |
| **Dual combination, n (%)**  |  |  |
|  ERA+PDE5-I | 68 (17) | 102 (36) |
|  PDE5-I+prostanoid | 2 (1) | 16 (6) |
|  ERA+prostanoid | 31 (8) | 3 (1) |
| **Triple combination, n (%)** | 0 (0) | 5 (2) |
| **Calcium channel blockers, n (%)** | 5 (1) | 2 (1) |
| **No treatment, n (%)** | 28 (7) | 3 (1) |

ERA: endothelin-receptor antagonist; PDE5-I: phosphodiesterase type-5 inhibitor

\*ERA treatment included 12 patients in PATENT Trial.

#PDE5-I group included 10 patients in SERAPHIN Trial.

¶Prostanoid treatment included 11 patients in UT Trial and 3 patients in GRIPHON Trial.

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| Supplementary Table S2. Characteristics of IPAH patients in baseline risk assessment according to simplified version of 2015 ESC/ERS risk stratification |
|  | **Low risk** | **Intermediate risk** | **High risk** | **All** |
| Subjects, n (%) | 76 (24) | 207 (64) | 39 (12) | 322 |
| Age, years | 36±15 | 42±16 | 39±14 | 38±14 |
| Female, n (%) | 53 (70) | 143 (69) | 19 (49) | 215 (67) |
| WHO FC, n (%) |  |  |  |  |
| Class I-II | 55 (72) | 50 (24) | 0 (0) | 105 (33) |
| Class III | 21 (28) | 147 (71) | 31 (80) | 199 (60) |
| Class IV | 0 (0) | 10 (5) | 8 (21) | 18 (6) |
| 6MWD, meters | 421±104 | 363±94 | 257±100 | 369±107 |
| BNP, ng/L | 46 (23, 81) | 204 (132, 412) | 661 (370, 844) | 160 (71, 390) |
| NT-proBNP, ng/L | 171 (51, 287) | 1154 (560, 1918) | 2309 (2095, 2703) | 1679 (264, 1893) |
| Hemodynamics |
| RAP, mmHg | 5 (3, 6) | 8 (4, 12) | 15 (10, 22) | 7 (4, 11) |
| mPAP, mmHg | 62 (44, 65) | 62 (52, 70) | 61 (58, 81) | 62 (51, 78) |
| PAWP, mmHg | 7 (5, 10) | 7 (5, 10) | 9 (7, 11) | 9 (6, 11) |
| CI, L/min/m2 | 3.1 (2.7, 3.7) | 2.2 (2.0, 2.4) | 1.8 (1.6, 1.9) | 2.4 (2.0, 3.1) |
| PVR, Wood units | 10 (7, 12) | 15 (12, 18) | 19 (15, 26) | 14 (10, 17) |
| SVO2, % | 71 (66, 77) | 61 (57, 67) | 47 (44, 55) | 64 (57, 70) |
| Initial therapies (within 3 months after diagnosis), n (%) |
| No specific therapy | 5 (7) | 17 (8) | 1 (3) | 23 (7) |
| CCB therapy | 5 (7) | 0 (0) | 0 (0) | 5 (16) |
| Monotherapy | 55 (72) | 120 (58) | 26 (67) | 201 (62) |
| Combination therapy | 11 (15) | 70 (34) | 12 (31) | 93 (29) |

Values are expressed as mean±SD, medians (interquartile range) or n (%), unless otherwise stated. BMI: body mass index;BNP: brain natriuretic peptide; CCB: calcium channel blocker; CI: cardiac index; mPAP: mean pulmonary arterial pressure; 6MWD: 6-minute walking distance; NT-proBNP: N-terminal fragmental of pro-brain natriuretic peptide; PAWP: pulmonary artery wedge pressure; PE: pericardial effusion; PVR: pulmonary vascular resistance; RAP: right atrial pressure; SvO2: mixed venous oxygen saturation; WHO FC: World Health Organization functional class.

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| **Supplementary Table S3. Change in variable between baseline and re-evaluation** |
|  | **Baseline** | **Re-evaluation** | **p-value** |
| Subjects, n  | 392 | 286 |  |
| **Risk assessment, n (%)** | <0.001 |
| Low risk | 96 (25) | 85 (30) |  |
| Intermediate risk | 267 (68) | 159 (56) |  |
| High risk | 29 (7) | 42 (15) |  |
| **WHO FC, n (%)** | <0.001 |
| Class I-II | 132 (34) | 76 (27) |  |
| Class III | 234 (60) | 122 (43) |  |
| Class IV | 26 (7) | 25 (9) |  |
| 6MWD, meter | 379±107 | 396±120 | 0.009 |
| BNP, ng/L | 211 (65, 426) | 184 (64, 453) | 0.698 |
| NT-proBNP, ng/L | 748 (255, 1679) | 806 (146, 2326) | 0.098 |
| **Haemodynamics\*** |
| RAP, mmHg | 6 (3, 10) | 6 (4, 11) | 0.073 |
| mPAP, mmHg | 58 (48, 68) | 57 (40, 65) | 0.068 |
| PAWP, mmHg | 8 (6, 10) | 10 (7, 11) | 0.069 |
| CI, L/min/m2 | 2.4 (1.9, 3.0) | 2.6 (2.2, 3.4) | 0.009 |
| PVR, Wood units | 14 (9, 18) | 10 (5, 15) | 0.004 |
| SVO2, % | 62 (56, 69) | 65 (58, 74) | 0.038 |
| **Imaging (echocardiography)** |
| RA area, cm2 | 22 (16, 30) | 23 (17, 34) | 0.224 |
| Pericardial effusion |  |  | <0.001 |
| No PE, n (%) | 260 (66) | 170 (67)  |  |
| Minimal PE, n (%) | 89 (25) | 73 (29) |  |
| PE, n (%) | 8 (2) | 10 (4) |  |
| **Cardio-pulmonary exercise testing** |
| Peak VO2, mL/min/kg | 14±4 | 13±4 | 0.668 |
| VE/VCO2 slope | 56±33 | 52±28 | 0.108 |
| **Therapies, n (%)** | <0.001 |
| No specific/CCB therapy | 33 (8) | 5 (2) |  |
| Monotherapy | 258 (66) | 155 (54) |  |
| Combination therapy | 101 (26) | 126 (44) |  |

\*Haemodynamics of re-evaluation could be successfully measured in 44 patients.

Values are expressed as mean±SD, medians (interquartile range) or n (%), unless otherwise stated. BNP: brain natriuretic peptide; CCB: calcium channel blocker; CI: cardiac index; mPAP: mean pulmonary arterial pressure; 6MWD: 6-minute walking distance; NT-proBNP: N-terminal fragmental of pro-brain natriuretic peptide; PAWP: pulmonary artery wedge pressure; PE: pericardial effusion; PVR: pulmonary vascular resistance; RA: right atrium; RAP: right atrial pressure; SvO2: mixed venous oxygen saturation; VE/VCO2: ventilatory equivalents for carbon dioxide; VO2: oxygen consumption; WHO FC: World Health Organization functional class.