2007 7

[1]

2003 3

pulmonary arterial hypertension PAH

11

PAH CAV-1 KCNK3 TBX4 EIF2AK4 BMP9 [2-6]

PAH 2007

PAH

2007

PAH [7]

chronic thromboembolic pulmonary hypertension, CTEPH [8]

CTEPH

optical coherence tomography, OCT

/ Pd/Pa

[9] CTEPH

CTEPH

EARLY

World Health Organization, WHO

[10]

REVEAL PAH

[11]

AMBITION

[12]

PAH

11 4 5 6

2008 2013 2018

2016 ESC

ERS

[7]

2007

11

2007

11 2007

/ 1¨

/ / arterial hypertension, IPAH

PAH

A

B

C /

pulmonary hypertension pulmonary arterial hypertension idiopathic pulmonary arterial hypertension

pulmonary hypertension pulmonary arterial hypertension

2007

1

pulmonary embolism

[13]

2

pulmonary hypertension

1

mean pulmonary artery pressure, mPAP ≥25 mmHg

1 mmHg=0.133 kPa [7] mPAP 14±3 mmHg 20 mmHg

PAH 2018 6

2

PAH

mPAP≥25 mmHg pulmonary 1% > artery wedge pressure, PAWP ≤15 mmHg 65 10% >3 Wood [7] /

idiopathic pulmonary [14] 80%

	mPAP≥25 mmHg
	mPAP≥25 mmHg PAWP≤15 mmHg PVR>3 Wood
	/
	mPAP≥25 mmHg PAWP>15 mmHg
	DPG<7 mmHg / PVR≤3 Wood
	DPG≥7 mmHg / PVR>3 Wood

mPAP PAWP PVR DPG dPAP DPG=dPAP-PAWP 1 mmHg=0.133 kPa

万方数据

1.1 PAH	3.1
1.2 PAH	3.2
1.3 PAH	3.3 /
1.4 PAH	3.4
1.5 PAH	3.5
1.5.1	
1.5.2 HIV	4.1 CTEPH
1.5.3	4.2
1.5.4	4.2.1
1.5.5	4.2.2
1.6 PVOD / PCH	4.2.3
1.7 PPHN	4.2.4
	4.2.5
	4.2.6
2.1 HFpEF	5.1
2.2 HFrEF	5.2
2.3	5.3
2.4	

PAH 2011 IPAH 1 3 5~10/ 15~60/ 92.1% 75.1% [15] IPAH PAH [22] PAH PAH PAH 2/3[16] IPAH 50~65 PAH 20 80 36 [17] 3[7] PAH 1 3 5 68% 48% 34%[18] 20 90 PAH 2006 IPAH PAH PAH 1 89.3%[16] 2010 IPAH PAH PAH 1 2 3 89% 68% 55%[19]

PAH PAH [20] IPAH PAH PAH

IPAH [21] PAH / 2006 PAH IPAH PAH 1 3 5 68.0% / 38.9% 20.8%[21] 2007

PAH

DNA

RNA

[23] [

nitric oxide, NO

][24-25]

[MAPK

Rho/ROCK PI3K/AKT

BMP / β TGF-β

κB NF-κB Notch][22-23 26-27]

PAH

PAH IPAH

PAH 9

BMPR2 BMP9 ACVRL1 ENG

SMAD9 BMPR1B TBX4 CAV1 KCNK3

50%~80% PAH 20%~50%

IPAH [7,28]

BMPR2 PAH IPAH

70%~80% PAH

20%~40% IPAH BMPR2

[29] BMPR2 PAH

IPAH 53% 15%[30] BMPR2

PAH

PAH BMPR2 7.5%

PAH BMPR2

12.3%[31] BMPR2

PAH 20%

14%

42%

[28] BMPR2

IPAH/ PAH

[29-30]

BMP9 IPAH [6] BMP9

IPAH 22 6.7%

IPAH BMP9

BMPR2 2 IPAH

PAH

ACVRL1 ENG

71%

[32]

PAH

IPAH

PAH PAH

PAH

PVOD PCH

EIF2AK4 [5]

PVOD/PCH 9%~25% EIF2AK4

[33]

PVOD/PCH

EIF2AK4

PVOD/PCH PVOD/PCH

Sanger

PAH <500 μm

PAH

PAH

PAH PVOD/PCH /

CTEPH

[34-35] CTEPH

[36]

万方数据

38%

PAH

C

[D-
B BNP N B
NT-proBNP]
C

X CT
HIV
C

1/5
2 IPAH
WHO ~

PAH
CTEPH
CTEPH

1.

S >0.21 mV PAH
89% 81%[37] QRS ≥120 ms IPAH
[38]

PAH 5 25.4%

万方数据

[39] PAH ≤2.8 m/s

a C

2. X PAH X ≤

2.8 m/s

2.9~3.3 m/s

a C

2.9~

3.3 m/s

≥3.4 m/s

X C

4.

3. PAH

[40]

3 1

[43-44] IPAH

DLco <45%

/ >1

[45]

>1.1 >25 mm PVOD/PCH IPAH

>21 mm <50% DLco

>18 cm^2

<105 ms /

>2.2 m/s 2 PAH

PAH

/

IPAH

PAH

3 [46]

TAPSE Tei

[41-42]

5.

CTEPH CT

[47] PAH PVOD/PCH

6. CT CT

[48]

CT

≥29 mm / ≥

万方数据

1.0 3~4 / >1 CT PVOD/PCH PVOD CT PCH CT 1/3 PAH CT / CTEPH CTEPH CT CT CT [49] CT CT

7. [7] [50] IPAH

8. [51]

9.

10. IPAH Abernethy CT

11.

1

3

eV

eNqr c dianq r nc

1

PAH

PAH [59-63] OCT

5 mm

IVUS

C

IPAH PAH PAH

CTEPH OCT

C

PB/U c IPAH

C PAH

3 IVUS OCT

C

IPAH PAH

PAH PAH

C

CT

/

30°~60°

30°~60°

30°~60° 30°~

60°

[58]

intravenous

ultrasound, IVUS optical

coherence tomography, OCT

CT

a C

CTEPH a C

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6

5

3~6

[72]

PAH PAH

PAH

C

PAH 3~6

1 C

PAH

C

PAH

a C

1.

IPAH

[73]

2.

[74]

3.

4.

5.

/

6. 1/4

<85%

WHO <

60 mmHg

1 500~

2 000 m

1. CTEPH

IPAH PAH PAH

[75]

2.

50% PAH [12] PAH NT-proBNP [83] 1.6%

SERAPHIN PAH [84] PAH / [85]

2. 5

5 5 cGMP NO/cGMP 5 PAH 5 [86-87] PAH 6 min [88] SUPER-2 3 60% 46% 6 min [89] PAH [90-92] 5 5

AMBITION PAH [10] 5 PAH

EVALUATION PAH PAH [95-96]

PAH 5 PAH PAH 5

3.

NO cGMP PAH CTEPH PATENT-1 PAH 6 min NT-proBNP [97] PATENT-2 PATENT-1 1 [98] PAH[99] PAH[100] PAH 2 93%[99] 49% 9% 6% 11% 5

4.

cAMP

PAH PAH PAH [93-94] PAH 3~5 min

万方数据

WHO PAH

[101-103]

PAH

70%[104]

2~4 ng · kg

PAH

PAH 62.5~125 mg 2 2 mg·kg^{-1}·d^{-1} 2

PAH 5~10 mg 1 1.25 ~2.5 mg 1

PAH 10 mg 1

5

20~80 mg 3 <1 0.5~1 mg·kg^{-1}·d^{-1} 3 <20 kg, 10 mg 3 > 20 kg, 20 mg 3

40 mg 1 10~20 mg 1 2.5~10 mg 1

5~10 mg 2 1.25~2.5 mg 2

PAH CTEPH 1 mg 3 2.5 mg 3

PAH 2~4 ng·kg^{-1}·min^{-1} 20~40 ng·kg^{-1}·min^{-1} 100 ng·kg^{-1}·min^{-1}

PAH

PAH

IP

PAH

<60%

PAH CTEPH

PAH

PAH WHO

				WHO			
CCB			C		C	-	-
ERA							
			A		A	b	C
			A		A	b	C
			B		B	b	C
PDE-5i							
			A		A	b	C
			B		B	b	C
		b	B	b	B	b	C
sGC							
			B		B	b	C
PGI_2							
	a	-	-		A		A
		-	-		B	b	C
		-	-	a	C	b	C
		-	-		B	b	C
		-	-	b	B	-	-
IP							
		I	B		B	-	-

PAH WHO CCB ERA PDE-5i 5 sGC

PGI_2 a -

PAH WHO

	WHO			
+	B	B	b	C

[114-115]

>20 mmHg

<85%

PAH WHO

ECMO

[116]

ECMO

<2 L · min^{-1} · m^{-2}

6 min <350 m

[117-118]

PAH

PVOD PCH

万方数据

IPAH 3 23%
9%
1/3 PAH

[119] ECMO PAH
3 1 5
93% 90% 87%[120] PAH
18 IPAH
1 3 77.8%
72.2%[121]

[122-123]

PAH
9
WHO ICU

>110 /min
<90 mmHg

132 PAH 8%
[124] 231 PAH

CTEPH 6

2.8%[125] IPAH
6
15.8%[38]

PAH CTEPH

1%~6%[7]

CT

PAH
1. PAH PAH

PAH 5%~10%[126]

PAH PAH

2. PAH

4

1

2

3 PAH

IPAH

4 PAH

PAH

PAH

3. PAH

PAH

/

PAH

[7,127-128]

[129]

PAH

PAH PAH PAH

PAH

PAH

PAH

1

2

3

PAH 1

2. PAH

PAH

/

ESSDAI

PAH

PAH

PAH

3. PAH

PAH

1

physician global assessment PGA)<1

2 PAH

2015 CSTAR

/

PAH [139]

PAH

PAH

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Child-Pugh

B C

β

4.

IPAH REVEAL

2 5

67% 40%[144]

PAH

2

10% 5%

PAH[145] PAH

[7] PAH

PAH

PAH 3

85.9%[146]

PAH PAH

[147]

10 PAH

[148]

HIV

HIV PAH HIV

PAH 0.46%

IPAH 2 500 PAH

HIV PAH PAH 2%~

7%[149] HIV HIV

PAH 1995 0.21% 2006

0.03% [150

>3
362 PAH 88% IPAH/
PAH PAH
57% 36%[159]
10
IPAH/ PAH 2
5 90% 75%[158]

WHO
2011
PVRI
10 [160] IPAH
PAH PAH

2.

[167]

PAWP HFpEF
PAWP <15 mmHg[168]

β

CTEPH [179-180] 3 2.2%~3.5%
CT
5 82% 10 75%[183]
PAH 3
CTEPH

CTEPH 10 CTEPH

[184-186]

CTEPH

0 5 95%

CTEPH PAH

PAH

/

CTEPH [8]
MERIT PAH
CTEPH
PAH
CTEPH [181]
CTEPH

CTEPH

CTEPH 56.8%
[182]
1

2

3

万方数据

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RAP, mmHg		
RVP, mmHg		
PAP, mmHg		
PAWP, mmHg		
CO L/min		
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PVR Wood		
TPR Wood		
SVR Wood		

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