

Severe Idiopathic Pulmonary Arterial Hypertension(IPAH-s) During Pregnancy (13 Cases)

Jun Zhang, MD

Xiaohui Xu, Yanna Li, Hong Gu, Liyun Zhao, Jinglan Zhang



**Transfer diagnostic center of heart disease in pregnancy
Obstetrics and Gynecology medical center of severe
Cardiovascular**

**Beijing Anzhen Hospital
Capital Medical University
China , Beijing**

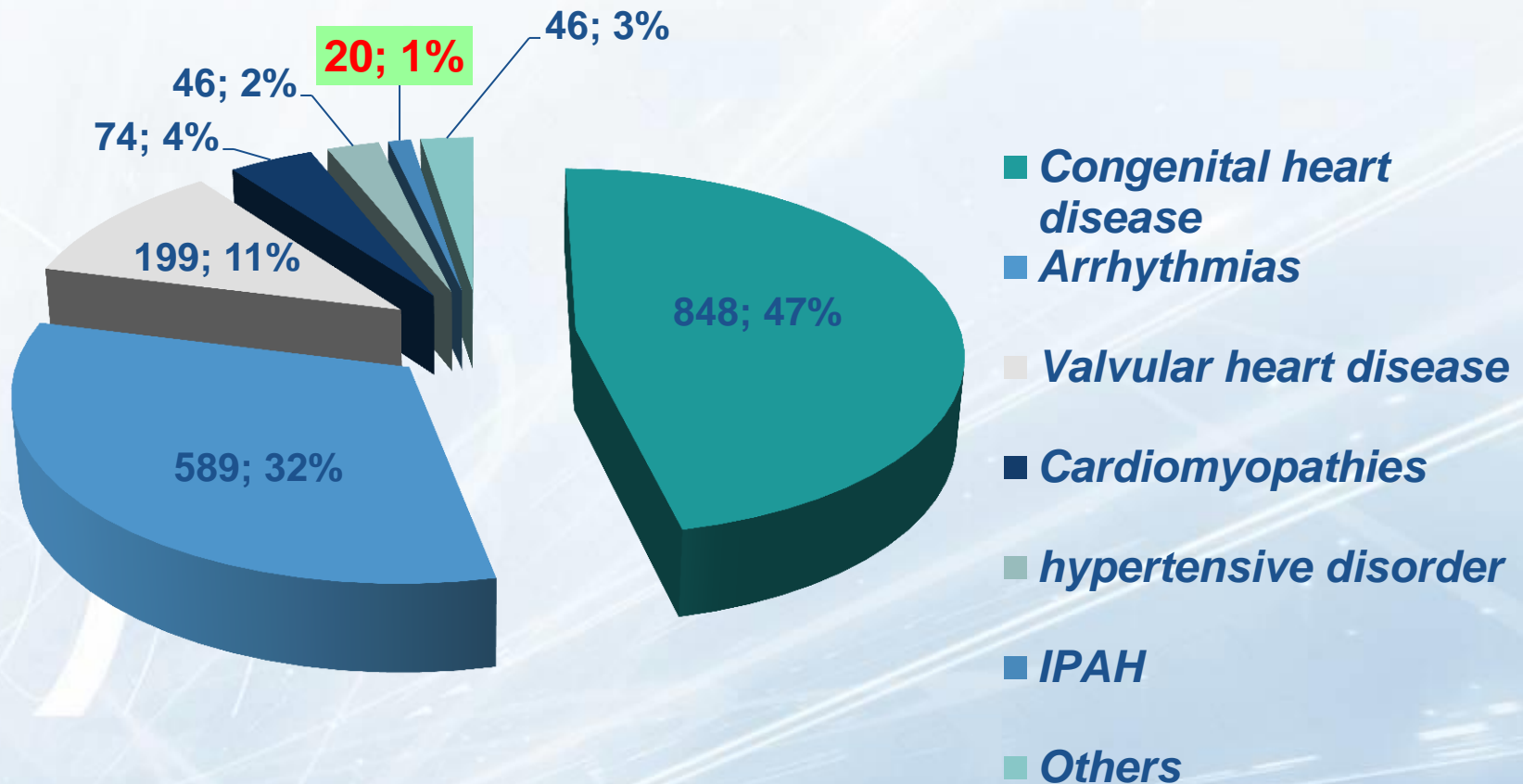


Presentation outline

- ❖ The evidence or experience about IPAH in pregnancy
- ❖ What is the risk for pregnancy?
- ❖ What are the causes of death and the inducing factors?
- ❖ How do our team deal with these patients?
- ❖ Our experience



Heat disease classification in P-

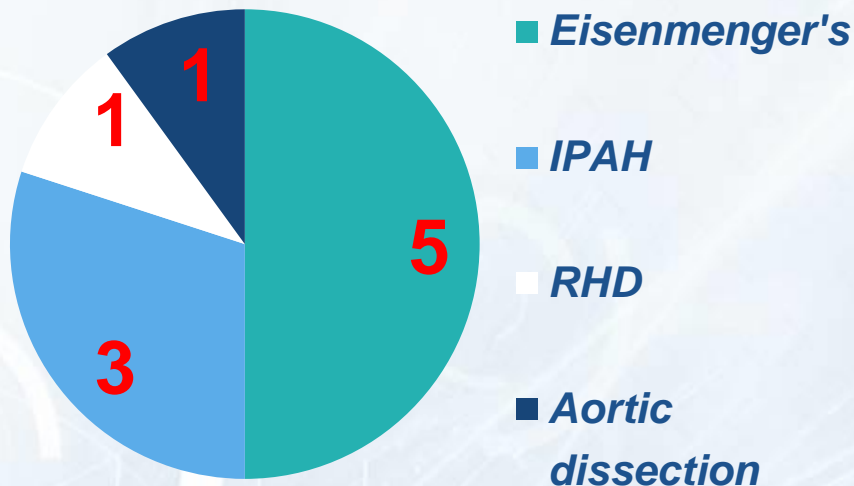


Mar, 2007~Nov, 2015, Beijing Anzhen Hospital



Constituent ratio

10 death cases



PH

✓ The percentage of IPAH in PH cases in our study is 4.9%(20/409)

✓ The maternal mortality is 23% in pregnancy with IPAH

Mar, 2007~Nov, 2015, Beijing Anzhen Hospital



Baseline characteristics of patients with severe IPAH

Case	Age (y)	BMI	GP	Diagnosis made (wk)	NYHA	sPAP* (mmHg)	EF** (%)	BNP (pg/ml)
1	29	26	G1P0	26	IV	91	67	203
2	25	23.8	G4P1	32	III	71	75	1657.4
3	26	22	G2P1	30	III	107	69	1710.2
4	22	21.6	G0P0	20	III	96	72	196.8
5	22	25.9	G1P0	35	III	102	43	2608.5
6	28	24	G1P0	39	III	74	74	227.2
7	25	20.8	G0P0	27	III	104	70	373.5
8	37	30.9	G3P1	30	III	100	73	435
9	41	25.4	G4P2	31	IV	106	75	180
10	27	22.1	G1P0	27	IV	105	60	648
11	27	21.1	G1P1	16	III	120	70	355
12	26	24.6	G0P0	19	III	102	64	209
13	22	26.9	G2P0	27	IV	107	62	1045

*=at the time when IPAH was firstly diagnosed; **= measured by echocardiograph



Baseline Characteristics of Patients with Severe IPAH

Case	Age (y)	BMI	GP	Diagnosis made (y)	NYHA	sPAP* (mmHg)	EF** (%)	BNP (pg/ml)
1	29	26	G1P0				67	
2	25	23.8	G4P1				75	
3	26	22	G2P1				69	
4	22	21.6	G0P0				72	
5	22	25.9	G1P0				43	
6	28	24	G1P0				74	
7	25	20.8	G0P0				70	
8	37	30.9	G3P1				73	
9	41	25.4	G4P2				75	
10	27	22.1	G1P0				60	
11	27	21.1	G1P1				70	
12	26	24.6	G0P0				64	
13	22	26.9	G2P0				62	1045

first diagnosis at the second and last trimester of pregnancy

heart failure

sPAP > 70 mmHg

Severity BNP UP

*=at the time when IPAH was firstly diagnosed; **= measured by echocardiograph



Management

- ❖ Hospitalization:
- ❖ MDT:
 - ✓ Obstetrics/anesthesiology/cardiology/pulmonology/ICU /neonatology
- ❖ Supportive therapies
- ❖ PAH-targeted therapies:
 - ✓ Epo(Epoprostenol)/S(sildenafil)/Tre(Treprostinil)/Iip(Iloprost)
- ❖ Termination



Management and Pregnancy Outcome

case	Advanced therapy			Delivery management				Maternal outcome
	Initial	During delivery	Postdelivery	Delivery	Anesthetic	Oxytocin	Blood (ml)	
1	25wk, S, Ilp	-	S, Ilp	31wk, CS	CSE	-	200	Alive
2	32wk, S, Ilp	-	S, Epo	32wk, CS	CSE	-	150	Death D5
3	30wk, S	Epo	S, Epo	30wk, CS	CSE	5U	230	Alive
4	20wk, S	-	S, Epo	21wk, TA	CSE	-	100	Alive
5	37wk, S	-	Epo	37wk, CS	CSE	5U -	800	Death D2
6	None	-	S, Epo	39wk, CS	GA	-	300	Alive
7	27wk, S	-	S, Ilp	28wk, CS	CSE	-	100	Alive
8	30wk, S	Epo	S, Epo	31wk, CS	CSE	-	200	Alive
9	31wk, S	-	S, Epo, Ilp	32wk, CS	CSE	-	200	Alive
10	27wk, S	Ilp	S, Epo, Ilp	28wk, CS	CSE	-	200	Death D3
11	16wk, S, Tre	Tre, Ilp	Tre	16wk, TA	CSE	-	100	Alive
12	22wk, S	-	S	24wk, TA	CSE	-	200	Alive
13	27wk, S, Ilp	Tre	Tre	28wk, CS	CSE	-	300	Alive

CS=cesarean section; CSE=continuous epidural anesthesia; Epo=Epoprostenol; GA= general anesthesia; Ilp=Iloprost; S=sildenafil; TA=therapeutic abortion; Tre=Treprostinil;

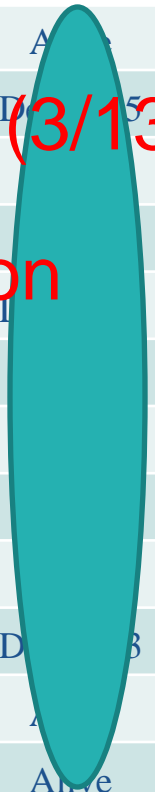


Management and Pregnancy Outcome

case	Advanced therapy			Delivery management/abortion				Maternal outcome
	Initial	During delivery	Postdelivery	Delivery	Anesthetic	Oxytocin	Blood (ml)	
1	25wk, S, Ilp	-	S, Ilp	31wk, CS	CSE	-	200	Alive
2	32wk, S, Ilp	-	S, Epo	32wk, CS	CSE	-	170	Deceased
3	30wk, S	Epo	S, Epo	30wk, CS	CSE	5U	230	Alive
4	20wk, S	-	S, Epo	21wk, TA	CSE	-	100	Alive
5	37wk, S	-	Epo	37wk, CS	CSE	5U	800	Deceased
6	None	-	S, Epo	39wk, CS	GA	-	300	Alive
7	27wk, S	-	S, Ilp	28wk, CS	CSE	-	100	Alive
8	30wk, S	Epo	S, Epo	31wk, CS	CSE	-	200	Alive
9	31wk, S	-	S, Epo, Ilp	32wk, CS	CSE	-	200	Alive
10	27wk, S	Ilp	S, Epo, Ilp	28wk, CS	CSE	-	200	Deceased
11	16wk, S, Tre	Tre, Ilp	Tre	16wk, TA	CSE	-	100	Alive
12	22wk, S	-	S	24wk, TA	CSE	-	200	Alive
13	27wk, S, Ilp	Tre	Tre	28wk, CS	CSE	-	300	Alive

maternal mortality is 23% (3/13)

3 therapeutic abortion
10 newborn alive



Death Cases

❖ Causes of death : pulmonary hypertensive crises, refractory heart failure

❖ Inducing factors:

❖ Key point:

Anesthesia effect

Delivery of fetal and placenta

Bleeding

(In our study three death cases occurred PH crisis just at the moment of expelling the baby and placenta)

Hemodynamic instability

Pulmonary hypertensive crises

Vicious spiral

heart failure

hyoxemia

acidosis

emergency orotracheal intubation

drug resistance

bleeding , etc



Our experience

- ❖ Severe IPAHA is the contraindication to pregnancy, but how to diagnose before pregnancy still is a problem
- ❖ The details and MDT is the key of success
 - ✓ Routinely use of Esmarch rubber bandages
 - ✓ If removal the placental ,5-10 minutes should be delayed after the childbith or naturally
 - ✓ Oxytocin should be avoid, intrauterine packing gauze or balloon can be used
 - ✓ Preventing postpartum hemorrhage
 - ✓ The individualized application of the vasoactive drugs



I thank You!

Dr. JUN ZHANG

E-Mail: drzhangj@outlook.com

