

SURVEY

ACUTE VASOREACTIVITY TESTING in PEDIATRIC IDIOPATHIC OR FAMILIAL PAH

Q1. WHAT DO YOU USE FOR THE AVT?

1. Inhaled Nitric Oxide
2. Nitric Oxide+Oxygen (specify FiO₂ and PPm of NO)
3. 100% Oxygen
4. iv Epoprostenol
5. Inhaled Iloprost
6. Others (specify)

Q2. THE PATIENT CONDITION DURING THE CATHETERIZATION IS...

1. General Anesthesia & Mechanical Ventilation
2. Sedation in spontaneous breathing
3. Awake patient
4. The condition is not always detailed in the cath report

Q3.WHICH CRITERIA DO YOU USE TO CONSIDER A +POSITIVE AVT

1. Sitbon criteria
2. Barst criteria
3. almost normalization of PAP and PVR < 4.5 WU.m²
4. Others (specify)

Q4. FOR PATIENTS WHITH +AVRT in FC I-II YOUR FIRST LINE THERAPHY IS:

1. Calcium channel blockers monotherapy
2. Calcium channel blockers+ oral drug (ERA/5PDEi)
3. other (specify)

Q5. FOR PATIENTS WHITH +AVRT in FC III YOUR FIRST LINE THERAPHY IS:

1. Calcium channel blockers monotherapy
2. Calcium channel blockers+ oral drug (ERA/5PDEi)
3. Calcium channel blockers+ Prostacyclin

4. Calcium channel blockers+ Prostacyclin + oral drug (ERA/5PDEi)
5. ERA/ 5PDEi
6. Combination Therapy not including CCB

Q6. FOR A PATIENT WITH + POSITIVE AVT , TREATED WITH CALCIUM CHANNEL MONOTHERAPY , YOU REPEAT THE CATH AFTER:

1. 6 months of therapy
2. 12 months
3. 3 months
4. If the patient is doing well, and the echo shows improvement, I don't repeat the cath

Q7. WHAT DO YOU CONSIDER "A MAINTAINED GOOD RESPONSE" IN THE LONG TERM AFTER CALCIUM CHANNEL BLOCKER THERAPY:

1. Patient remains on FC I-II , and event –free, in spite of high pressure and resistances in the catheterization.
2. Remains on FC I/II and there is evidence of almost normalization of PA and PVR
3. Other. Specify)

Q8. IF AFTER 6 MONTHS OF CALCIUM CHANNEL BLOCKER MONOTHERAPY the PVR/mPAP HAVENT CHANGED FROM BASAL , YOU:

1. Remove the Calcium Channel and Start an ERA/PDEi
2. Add Another Therapy to the Calcium Channel Blocker
3. Depending on The PVR at the Second Catheterization

Q9. HOW MANY OF THE PEDIATRIC PATIENTS WITH IDIOPATHIC PAH SEEN IN YOUR INSTITUTION (OR IN YOUR EXPERIENCE IN DIFFERENT INSTITUTIONS) HAVE HAD A +AVRT AND MAINTAINED NORMALIZATION OF PAP IN THE LONG TERM (MORE THAN 5-10 YEARS) WITH CCB? Example : 2 of 10 (20%)

Q10. DO YOU PERFORM ALSO AVRT IN OTHER TYPES OF PH? (e.: postoperative, associated to lung disease, etc...)

1. yes, always, in every PH patient that is catheterized
2. Only in PAH patients
3. Only in Idiopathic or familial PAH.

Q11. DO YOU ALSO PERFORM AVRT IN THE NEXT CATHETERIZATIONS OF YOUR PATIENTS DURING FOLLOW UP?

1. Yes, always
2. Never

3. Depending on the case.

Name:

Institution: