

Pulmonary Endarterectomy: the Best Treatment Option of CTEPH

▲ Yanhong NIU from MDWEEKLY

“Pulmonary endarterectomy is the best treatment option of chronic thromboembolic pulmonary hypertension (CTEPH) and should always be the first line of treatment.” Prof Michael M Madani concluded in his presentation.

Madani pointed out that surgery by means of a complete pulmonary thromboendarterectomy (PTE) is the best option of treating CTEPH. However, surgery is technically challenging, and excellent outcomes require access to an expert center. Besides, operability determination challenges, and access to an expert center may be limited in certain areas of the world.

As for operability, it is influenced by imaging, surgical experience, hemodynamics and patient factors. Pepke Zaba J et al investigated the clinical characteristics and current management of patients enrolled in an international CTEPH registry. The results showed that not all patients were operable; however the evaluation of operability was partly subjective and

depended from center size, small centers select easier cases; effect of center expertise or selection biased in surgical centers. The reasons why patients were assessed as nonoperable were mainly clot inaccessibility, comorbidities, PH severity and morphological lesions imbalance, PVR > 1500 dsc-5 and age.

Who is not a candidate? The influencing factors include: operability; surgical experience; center's experience; patient factors such as age, comorbidities, and technical difficulties; hemodynamic versus anatomical obstruction.

Pretorius GD, et al retrospective reviewed 1547 cases of PEA performed. He found that age over 80 was not a contraindication to surgery and these patients had significant benefit from surgery.

Auger W found that patients with type III disease achieved a significant reduction in mPAP and PVR, even in those with PVR >1000, with similar perioperative mortality; Although the incidence of PVR > 400 postop was higher, but the clinical

significance of this difference between the groups is likely small.

As regard for truly inoperable patients, available options include second opinion from an expert center, medical therapy, transplantation, and balloon angioplasty when appropriate.

Medical treatment may play a role in inoperable and postoperative PH, but there is only one approved drug. And the effect on the obstructed vessels is uncertain. The CHEST study evaluated the efficacy of Riociguat in the treatment of patients with inoperable CTEPH or persistent/recurrent PH after PEA. The result showed that Riociguat significantly improved exercise capacity and pulmonary vascular resistance in patients with CTEPH.

In conclusion, CTEPH is severely underdiagnosed/misdiagnosed and in many cases not referred. Surgery is the best treatment option and should always be the first line of treatment. Referral to an expert center should always be considered as a first option, and as a second opinion.



Medical therapy is available for inoperable and residual PH patients, but its role is still unclear.

Orphan Drug Clinical Trials: Gadgets Will Help!

▲ TaoWEN from MDWEEKLY



Due to insufficient number of patients, the efficacy of drug for rare diseases, which is defined as a condition or disease affecting fewer than 5 in 10,000 in EU, may be difficult to establish. How to deal this situation? “Creative thinking and adaptability” are the words uttered by Tom MacDonald, UK.

His first answer is “treatment allocation by minimization, not randomization.” That is to say, to minimize the standard deviation of the differences (SDD) between repeated measures in the same subjects.

“Patients are Precious.” He said, “so Multiple Measurements improve precision, reduce variance and increase power.”

For a parallel trial to detect a change of 10 mmHg Systolic blood pressure (SBP), we need 51 subjects using Clinic SBP. However, only 28 subjects are required by using ambulatory blood pressure monitoring (ABPM).

Even the Apple or Android cell phones with blood pressure monitor with bluetooth connection with cuff, and validated to ESH

standard are not ideal devices, not mention the ABPM used in the hospital. He claimed, repeated ‘measures’ is boring for patients. “We need to embrace new technology!”

He felt very excited that here comes the cuffless blood pressure monitoring device using a disposable patch with miniaturized electronics, which can measure blood pressure continuously and non-interferingly. Now patients can wear “blood pressure-sensing underpants.”

A series of implantable devices are on the way, including implantable ECG Monitoring, implantable PA pressure monitoring, implantable left atrial pressure monitoring, wearable continuous pulse

oximeter, finger photoplethsmography that can measure beat to beat BP, pulse, stroke volume, cardiac output and left ventricular end diastolic pressure, continuous glucose monitoring connected watch, thoracic bioimpedance monitoring, wearable, skin-like device 'monitors cardiovascular 24/7', tattoo device to monitor sweat sodium or lactate.

Now the era has come to “use high technology to capture large amounts of outcome data over time”, Tom stated.

He also gave some advices on the skills of improving the power, like studying more severe disease, carry-over effects, using expensive placebo and surrogate outcome measures, and so on.

