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John Conte
*Chest* 2004;126;63-71
DOI: 10.1378/chest.126.1_suppl.63S

This information is current as of April 22, 2006

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http://www.chestjournal.org/cgi/content/full/126/1_suppl/63S
Surgical Treatments/Interventions for Pulmonary Arterial Hypertension*

ACCP Evidence-Based Clinical Practice Guidelines

Ramona L. Doyle, MD, FCCP; Douglas McCrory, MD, MHSc; Richard N. Channick, MD, FCCP; Gerald Simonneau, MD; and John Conte, MD, FCCP

While considerable advances have been achieved in the medical treatment of pulmonary arterial hypertension (PAH) over the past decade, surgical and interventional approaches continue to have important roles in those patients for whom medical therapy is unavailable or has been unsuccessful. These techniques include pulmonary thromboendarterectomy for chronic thromboembolic pulmonary hypertension, thoracic transplantation, and atrial septostomy. This chapter will provide evidence-based recommendations for the selection and timing of surgical and interventional treatments of PAH for physicians involved in the care of these complex patients.

(CHEST 2004; 126:63S–71S)

Key words: atrial septostomy; balloon angioplasty; heart-lung transplantation; lung transplantation; pulmonary hypertension; pulmonary thromboembolic disease; pulmonary thromboendarterectomy; survival

Abbreviations: AS = atrial septostomy; BLT = bilateral lung transplantation; CO = cardiac output; CTEPH = chronic thromboembolic pulmonary hypertension; HLT = heart-lung transplantation; IPAH = idiopathic pulmonary arterial hypertension; ISHLT = International Society of Heart and Lung Transplantation; LT = lung transplantation; NYHA = New York Heart Association; OB = obliterative bronchiolitis; PAH = pulmonary arterial hypertension; PAP = pulmonary artery pressure; PH = pulmonary hypertension; PTE = pulmonary thromboendarterectomy; RAP = right atrial pressure; SLT = single lung transplantation; V˙/Q˙ = ventilation/perfusion

Recent developments in the medical treatment of pulmonary arterial hypertension (PAH) have overshadowed the role of interventions in the treatment of PAH. In 1981, the first heart-lung transplantation (HLT) in the world was performed in a patient with primary pulmonary hypertension,1 but since that time the use of transplantation in patients with PAH has decreased.2 The use of prostanoid-based therapies is likely responsible for this decline in the use of transplantation, and the development of new classes of medications for PAH may impact the use of surgical and other interventional therapies for PAH in the future. The rationale for surgical/interventional therapies in PAH is based on the identification of subsets of patients in whom a specific procedure is indicated. In the case of some procedures, such as atrial septostomy (AS), the treatment may merely serve as palliation or as a bridge to more definitive treatment, such as lung transplantation. Pulmonary thromboendarterectomy (PTE), however, offers a potential surgical cure for a subset of patients with chronic thromboembolic PAH (CTEPH). In this section, we examine the data for AS, PTE, and lung transplantation (LT) or HLT in the treatment of PAH.

As the basis for our recommendations, we considered studies with at least 10 subjects conducted in any patients with known or suspected idiopathic PAH (IPAH), PAH associated with congenital heart disease, and PH associated with chronic thromboembolic disease. We excluded studies in patients with PH associated with left-sided cardiac disease, such as congestive heart failure or valvular disease, as well as PH associated with high altitude. We excluded studies or analysis of studies of corrective surgical procedures for congenital heart disease-associated PH. Although we have attempted to provide an evidence-based overview of the available

*From Stanford University (Dr. Doyle), Stanford, CA; University of California, San Diego (Dr. Channick), San Diego, CA; Duke University Medical Center (Dr. McCrory), Durham, NC; Hôpital Antoine Béclère (Dr. Simonneau), Clamart, France; and Johns Hopkins University (Dr. Conte), Baltimore, MD.

For financial disclosure information see page 1S.

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Correspondence to: Ramona L. Doyle, MD, FCCP, Pulmonary And Critical Care Medicine, H3147 Stanford University School Of Medicine, Palo Alto, CA 94305; e-mail: rldoyle@stanford.edu

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data on AS, PTE, and transplantation, there are important limitations of the evidence due in large part to the quality and number of the studies forming the basis of our recommendations. When specific data were not available, we based our recommendations on expert opinion from centers that treat large numbers of patients with PH and have extensive experience in the procedures examined.

Atrial Septostomy (AS)

Patients with IPAH and a patent foramen ovale who were awaiting transplantation were found to have a survival advantage over those without a patent foramen ovale, suggesting that the creation of an intra-atrial right-to-left shunt could decompress the right ventricle and increase left ventricular preload, thereby increasing systemic blood flow and improving systemic oxygen transport despite arterial oxygen desaturation. Ideally, the impact of a decline in arterial oxygenation would be further limited by compensatory polycythemia. AS has been used as a palliative treatment in patients with advanced PH based on its potential to decompress the failing right ventricle and increase cardiac index. We identified four reports meeting our inclusion criteria. AS is indicated in severely symptomatic patients with PAH who are unresponsive to medical treatment. Limited reports of selection criteria for AS suggest that only patients with “severe PAH” should be considered for the procedure, although definitions of severe vary and include the following: New York Heart Association (NYHA) class III or IV, a history of recurrent syncope, or “refractory” right-heart failure. Improvements in cardiac index immediately following the procedure range from 15 to 58%. Arterial oxygenation decreases significantly immediately after the procedure. Most studies have shown very little immediate effect of AS on mean pulmonary arterial pressure (PAP). Hemodynamic improvements, however, have been shown to correlate with improved NYHA functional class and exercise tolerance as measured by 6-min walk distance. Quality-of-life data for AS in PAH are not available. Longer-term hemodynamics have been reported in only two studies. In one study, three NYHA class IV patients underwent repeat catheterization at 18 months, 21 months, and 27 months after AS, and a sustained decrement in mean right atrial pressure (RAP) and an increase in cardiac index were found. In another study, 4 of 15 patients who survived initial septostomy developed closure of their septal defect and underwent repeat AS, with 2 patients requiring more than one repeat procedure. In these limited reports of hemodynamics, no significant additional improvements in PAP or cardiac index were reported; in one study, RAP, which had remained unchanged immediately after septostomy, fell significantly at follow-up catheterization 7 to 27 months after the procedure.

In our summary analysis, 7 of 55 patients undergoing AS died during or immediately after AS (13% mortality); 30-day survival was 82%. Procedural mortality from single center reports, however, ranges from 5 to 50%. Appropriate selection of patients for AS is clearly critical, but the published literature suggests broad, not specific, selection criteria. Patients with the most advanced PAH, defined by markedly elevated pulmonary vascular resistance, arterial oxygen saturations < 80% at rest, and severe right-heart failure (manifested by low cardiac output [CO] and high RAP) appear more likely to die or worsen with AS.

This procedure should only be performed in institutions with an established record in the medical and surgical treatment of PAH, and where septostomy is performed routinely with low morbidity. In the absence of further research and innovation, our recommendation is that AS should be performed in patients with severe PAH in whom other therapies are either unavailable or have failed, and who find themselves in need of palliative or bridging maneuvers.

Recommendations

1. In select patients with PAH unresponsive to medical management, AS should be considered. Quality of evidence: low; net benefit: intermediate; strength of recommendation: C.

2. In patients with PAH, AS should be performed only at institutions with significant procedural and clinical experience. Quality of evidence: expert opinion; net benefit: substantial; strength of recommendation: E/A.

Pulmonary Thromboendarterectomy (PTE)

PTE may provide a potential surgical cure for PAH resulting from chronic thromboembolic PAH (CTEPH) affecting central pulmonary arteries (main, lobar, segmental). PTE for treatment of CTEPH was first described > 30 years ago. Prior to this, CTEPH was an autopsy finding. Since that time, nearly 1,000 PTE cases have been reported in the literature. As with many surgical procedures, published reports of PTE have not been well controlled. The PTE procedure must be distinguished from acute embolectomy for massive pulmonary embolism, not the topic of this review. PTE involves...
a true endarterectomy, resulting in dissection of well-organized thromboembolic material as well as part of the intimal layer of the pulmonary arterial bed. The accepted surgical technique has been described in detail previously.9,10 The procedure is currently performed via a median sternotomy with cardiopulmonary bypass, and involves profound induced hypothermia and periods of full circulatory arrest, during which time dissection of the thromboembolic material is accomplished. The goal of PTE is to reduce pulmonary artery pressures (PAPs), hopefully to normal, thereby leading to improved right ventricular function; in the ideal case, PTE surgery is completely curative.

**Diagnostic Tests To Determine Surgical Candidacy for PTE**

Ascertainment of the presence of surgically accessible thromboembolic disease is obviously critical to a successful hemodynamic outcome and this relies primarily on radiographic imaging techniques. The presence of concomitant microvasculopathy (small-vessel pulmonary vascular disease) or inaccessible distal disease may limit the response to PTE.

**Ventilation/Perfusion Scanning**

It is generally accepted that a normal perfusion scan (one with no perfusion defects) effectively rules out CTEPH as a diagnostic consideration, although the negative predictive value of a normal ventilation/perfusion (V/Q) scan finding is not known, as these patients are typically not offered surgery. In contrast, V/Q scans in patients with surgically accessible CTEPH typically demonstrate at least one segmental or larger mismatched perfusion defect,11 although one study12 reported that abnormalities seen on V/Q scan may underestimate the extent of disease as documented by pulmonary angiography.

**Chest CT Scanning**

There are several reports in the literature of helical CT scanning in the diagnosis of CTEPH. One report in 47 patients by Bergin et al13 found that CT scanning diagnosed central chronic thromboembolic disease with an accuracy of 0.79, comparable to angiography when surgical specimen was used as the "gold standard." However, in this study and others, there were patients in whom CT scanning missed the diagnosis of surgically accessible CTEPH. Therefore, it is generally believed that pulmonary angiography is required in most patients prior to surgical intervention. One indication for helical CT is in patients with unilateral obstruction in whom other diagnoses such as pulmonary artery sarcoma or fibrosing mediastinitis may be more common. One study14 found that in 27 patients with unilateral absent or diminished perfusion, CT scan more accurately (0.86) predicted the presence of another diagnosis that pulmonary angiography (0.69).

**Pulmonary Angiography**

Pulmonary angiography has been shown to be safe when performed by experienced operators.15 Although no systematic reports are available, it is generally agreed that pulmonary angiography is required to confirm surgical accessibility of chronic thromboemboli.

**Other Selection Criteria for PTE**

Data available on surgical selection of patients for PTE is limited, but we identified 21 reports meeting our inclusion criteria.8,16–30 While the selection of appropriate patients for PTE is critical in the success of the procedure, not all centers consistently report their selection criteria. The following basic criteria should be considered for selection of patients undergoing PTE: (1) NYHA functional class III or IV symptoms; (2) a preoperative pulmonary vascular resistance > 300 dyne·s·cm⁻⁵; (3) surgically accessible thrombus (in the main, lobar, or segmental pulmonary arteries), as determined by all appropriate radiographic studies; and (4) no severe comorbidities. In addition, some centers have suggested that the mean PAP preoperatively should be at least ≥ 40 mm Hg.

**Hemodynamic Outcomes Following PTE**

Sixteen, uncontrolled reports15–31 describe preoperative and postoperative hemodynamic results. Fifteen of these reports were limited to early (< 3 months) postoperative results. These reports confirm that PTE leads to significant reductions in PAP and improvement in CO. In most of the studies, the mean PAP was in the 45 to 50 mm Hg range preoperatively, decreasing to 25 to 30 mm Hg postoperatively. Studies variously report CO or cardiac index. In general, most reports indicated that preoperatively patients had cardiac indices in the range of 1.9 to 2.6 L/min/m².16,18–21 Postoperatively, cardiac index appeared to become near normal (confidence interval, 2.6 to 3.3 L/min/m²). It has been shown that some patients may have persistent PH despite PTE. However, no data regarding predictors of poor hemodynamic outcome are available.

**Functional Outcomes Following PTE**

Several reports14–19,21,22,28 we identified included some form of postoperative functional status out-
come. Most often, this was a description of changes in NYHA classification. These studies suggest that while the majority of patients are NYHA III or IV prior to PTE, approximately two thirds of patients surviving PTE report NYHA class I or II symptoms. In a single-center study of 306 patients, 73% of patients reported their dyspnea was “much improved” following PTE.\(^{14}\) Very little objective data, such as 6-min walk tests or formal exercise tests, are available in patients following PTE.

**Survival Following PTE**

Nineteen uncontrolled reports provide 30-day or “postoperative” mortality data, with mortality ranging from 7 to 35%. The 30-day mortality among the 802 patients reported in these 19 reports was 14% (691 surviving among 802 patients). Most early mortality is due to persistent PH, reperfusion pulmonary edema, sepsis, and hemorrhage. There are limited long-term mortality data available. Archibald et al\(^{14}\) reported 75% 6-year survival, with several very long-term (> 15 years) survivors. Although there are no controlled studies, the data support a survival benefit for PTE surgery, compared to historical controls.

**Alternatives or Adjuncts to PTE**

In patients who are deemed nonsurgical or high-risk surgical candidates, there are three potential options: medical therapy, balloon pulmonary angioplasty, and LT. Medical therapy may be considered for patients with CTEPH who have no surgical options (see section on Medical Therapy). Some authors have suggested a role for medical therapy for patients awaiting PTE. In one uncontrolled study, Nagaya and colleagues\(^{30}\) treated 12 patients with IV epoprostenol prior to the surgery and reported a 28% decrease in pulmonary vascular resistance. Two other uncontrolled studies\(^{37,38}\) have suggested a possible benefit to preoperative medical therapy in some patients with CTEPH, but without further controlled studies the benefit of medical therapy in this setting remains unclear. Feinstein and colleagues\(^{39}\) in a study of 18 patients, reported balloon pulmonary angioplasty for distal thromboembolism. Following the procedure, patients had significant improvements in 6-min walk distance (209 to 497 yards) and functional status with only one death. LT may also be a viable option for some patients with inoperable CTEPH. Although there are no formal studies evaluating their efficacy, inferior vena cava filter insertion and life-long anticoagulation is generally performed at most centers in all patients with CTEPH in order to reduce the risk of recurrent thromboembolism.

**Recommendations**

3. Patients with suspected CTEPH should be referred to centers experienced in the procedure for consideration of PTE. Level of evidence: expert opinion; benefit: substantial; grade of recommendation: E/A.

4. In patients with operable CTEPH, PTE is the treatment of choice for improved hemodynamics, functional status, and survival. Level of evidence: low; benefit: substantial; grade of recommendation: B.

5. In patients with CTEPH deemed inoperable or with significant residual postoperative PH, balloon dilation, PAH medical therapy, or LT may be considered. Level of evidence: low; benefit: small/weak; grade of recommendation: C.

**Lung Transplant and Heart-Lung Transplant (LT and HLT)**

LT has been a mainstay of treatment for PH since the 1980s. The first successful LT was a combined HLT performed in a woman with pulmonary vascular disease due to IPAH.\(^{1}\) Since that initial success, LT has been applied to a wide variety of conditions. Guidelines for the selection of candidates for LT have been previously published,\(^{40}\) and readers are referred to that publication for a more comprehensive review of the topic.

**Timing of Transplantation in PAH**

The timing of transplantation has been traditionally dependent on many factors. The primary diagnosis, stage of disease, response to conventional therapies, the availability of alternative or experimental treatments, and local waiting times for donors have all factored into the timing since the early 1980s. It was, however, the dramatic emergence, approval, and availability of medical therapies in the 1990s that changed the paradigm of evaluation and listing for most patients with PAH. Prior to these developments, the diagnosis of PAH mandated immediate listing and transplantation. Epoprostenol was the first therapy that could realistically be viewed as a bridge or alternative to transplantation for many patients with PAH.\(^{41}\) In the current era, LT in any form should not be considered until after the failure of medical therapy, factoring into account the time needed to complete a transplant evaluation and the time spent on the transplant list awaiting suitable organs. Death on the waiting list has been found to be a particular problem in patients with poor NYHA functional status. Patients who have NYHA class III...
or IV symptoms on presentation should be referred for LT evaluation while their response to therapy is being evaluated in order to avoid delays in evaluation and listing.

The findings of an elevated bilirubin, an elevated creatinine, and the pretransplantation requirement for supplemental oxygen have all been found by multivariate logistic regression analysis to be independent risk factors for mortality in the first year and at 5 years after LT. Unfortunately, these are commonly noted in patients with PH. In an International Society of Heart and Lung Transplantation (ISHLT) Registry subset analysis of patients transplanted between January 1995 and June 1997, donor and recipient age and a pretransplant pulmonary artery systolic pressure > 40 mm Hg were also associated with an increased 5-year mortality.

**Type of Transplant Procedure**

Transplant procedures currently being performed in patients with PAH include single LT (SLT), bilateral LT (BLT), and combined HLT. Patients with IPAH account for 5% of recipients registered in the ISHLT Registry. They represent 2% of SLT recipients, 8% of BLT recipients, and 24% of HLT recipients. Though not addressed in this review, PH may occur in patients with other primary lung diseases, and may also play a role in another 10 to 20% of recipients who received isolated LT.

There is significant variability among reports from individual transplant centers as to the type of procedure patients with PAH are likely to receive. Although HLT was the first successful procedure for PH and is still preferred at many centers, the majority of LT programs now include either SLT or BLT as options for patients with PAH. This shift has been based on both outcomes and pragmatic concerns, including a growing shortage of organs for transplantation. Proponents of SLT have argued that SLT is an easier procedure to perform, requires less time to complete, and results in less ischemic time and shorter cardiopulmonary bypass times than BLT or HLT. Detractors of SLT for PAH note the potential for V/Q mismatch and a higher likelihood of reperfusion injury. Proponents of BLT argue that BLT results in better hemodynamics, less V/Q mismatch, as well as fewer complications in the perioperative period, which might enable more “marginal lungs” to be utilized. By providing better overall lung function at the outset, BLT may also be more protective against the physiologic manifestations of chronic rejection. BLT may also offer a better long-term posttransplant survival than SLT. BLT, however, can be a more difficult procedure to perform, with the potential for increased morbidity due to longer ischemic and bypass times. HLT has the advantage of requiring only one airway anastomosis, a near absence of vascular complications, and the best hemodynamic outcomes. However, the donor organ shortage and the organ allocation system in the United States can prove disadvantageous to patients awaiting BLT, and especially those awaiting HLT. Thus, patients listed for HLT in the United States may have longer waiting times and their overall survival may be adversely impacted.

A survival advantage of BLT compared with SLT in IPAH has been reported, but in one study this was only true in patients with high preoperative PAP (> 40 mm Hg). Other centers, which have reported similar early complications following SLT, disagree about the impact of SLT vs DLT on overall mortality. A survival benefit of HLT over either SLT or DLT procedures has not been well elucidated, with some centers showing no difference in mortality, and others suggesting superior survival with the HLT procedure. There are no randomized studies to assess the relative efficacy of each transplant procedure in patients with PAH and, given the scarcity of organs, such information is unlikely to be available in the future. The optimal transplant procedure for patients with PAH will depend on individual patient characteristics and the availability of appropriate organs for transplantation. Decisions about which procedure is most appropriate for each individual patient should be left to centers with extensive experience in organ transplantation.

**Survival Following Transplantation**

The ISHLT database consisting of 13,453 transplant recipients shows that overall survival for SLT and BLT regardless of etiology of the lung disease is nearly equal up to 3 years after transplant. After that, however, the survival curves diverge and there appears to be a survival advantage for BLT. Early 30-day survival appears equivalent with either procedure in patients with IPAH as well, but there is a survival advantage with BLT for all time points up to 9 years that does not reach statistical significance. For BLT, the survival is 70%, 55%, 45% and 20%; for SLT, it is 65%, 50%, 40%, and 23% at 1 year, 3 years, 5 years, and 10 years, respectively. In the same database, survival following HLT is 65%, 45%, 40% and 25% at 1 year, 3 years, 5 years, and 10 years, respectively. For patients with IPAH, the survival following HLT is 70%, 50%, 40%, and 25% at 1 year, 3 years, 5 years, and 10 years, respectively. There does not appear to be a significant difference between HLT performed for congenital heart disease or IPAH. In the United Network for Organ Sharing...
database,\(^6\) survival in patients with PH following LT alone is 73% at 1 year and 56% at 3 years (no breakdown by SLT vs DLT is available). Survival following HLT is 76% at 1 year and 45% at 3 years for patients with IPAH; in patients with PAH associated with congenital heart disease, survival is 65% and 44%, respectively. The major limitation to long-term survival in LT is death due to chronic rejection, which is manifested in the lungs as obliterative bronchiolitis (OB). While any injury to the lung allograft (infection, rejection, ischemia, reperfusion injury) is likely to contribute to the development of OB, the relationship of OB to a recipient diagnosis of PH and the type of transplant procedure performed remains unknown.\(^4\),\(^6\)

**Hemodynamic Outcomes Following Transplantation**

Studies that have included postoperative hemodynamic data for PAH patients who underwent transplantation show that most of the patients had a preoperative mean PAP of 60 to 70 mm Hg, while postoperatively survivors had a mean PAP of 20 to 25 mm Hg. Other immediate hemodynamic improvements in cardiac index and pulmonary vascular resistance have also been documented. Direct comparison of hemodynamic outcomes in one study\(^7\) showed significant improvements in CO with both BLT and HLT, but the improvement in the HLT was significantly greater. In patients with PH undergoing transplantation, higher rates of reperfusion edema have been shown to occur in SLT vs DLT, which is thought to be due to the enhanced blood flow to the transplant allograft in a SLT. Increased blood flow and worse reperfusion injury have resulted in prolonged mechanical ventilation and protracted ICU stays in patients with PAH undergoing SLT.\(^4\),\(^7\)

**Functional Outcomes**

In terms of long-term functional status following LT or HLT, there are no data specific for patients with PAH; \(>80\)% of survivors of LT or HLT reported no limitation in activity at 1 year, 3 years, and 5 years following transplantation, and 40 to 50% were working either full-time or part-time.\(^2\)

**Recommendations**

6. **PAH patients with NYHA functional class III and IV symptoms should be referred to a transplant center for evaluation and listing for LT or HLT.** Level of evidence: low; benefit: substantial; grade of recommendation: B.

7. **Listed patients with PAH whose prognosis remains poor despite medical therapy should undergo LT or HLT.** Level of evidence: fair; benefit: substantial; grade of recommendation: A.

8. **In patients with PAH who are undergoing transplantation, the procedure of choice is BLT.** Level of evidence: low; benefit: intermediate; grade of recommendation: C.

9. **In children with PAH who are undergoing transplantation, the procedure of choice is BLT.** Level of evidence: low; benefit: substantial; grade of recommendation: B.

10. **In adult patients with PAH and simple congenital heart lesions, BLT with repair of the cardiac defect is the procedure of choice.** Level of evidence: low; benefit: intermediate; grade of recommendation: C.

11. **In adult patients with PAH and complex congenital heart disease who are undergoing transplantation, HLT is the procedure of choice.** Level of evidence: low; benefit: substantial; grade of recommendation: B.

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**Summary Of Recommendations**

1. **In select patients with PAH unresponsive to medical management, AS should be considered.** Quality of evidence: low; net benefit: intermediate; strength of recommendation: C.

2. **In patients with PAH, AS should be performed only at institutions with significant procedural and clinical experience.** Quality of evidence: expert opinion; net benefit: substantial; strength of recommendation: E/A.

3. **Patients with suspected CTEPH should be referred to centers experienced in the procedure for consideration of PTE.** Level of evidence: expert opinion; benefit: substantial; grade of recommendation: E/A.

4. **In patients with operable CTEPH, PTE is the treatment of choice for improved hemodynamics, functional status, and survival.** Level of evidence: low; benefit: substantial; grade of recommendation: B.

5. **In patients with CTEPH deemed inoperable or with significant residual postoperative PH, balloon dilation, PAH medical therapy, or LT may be considered.** Level of evidence: low; benefit: small/weak; grade of recommendation: C.

6. **PAH patients with NYHA functional class III and IV symptoms should be referred**
to a transplant center for evaluation and listing for LT or HLT. Level of evidence: low; benefit: substantial; grade of recommendation: B.
7. Listed patients with PAH whose prognosis remains poor despite medical therapy should undergo LT or HLT. Level of evidence: fair; benefit: substantial; grade of recommendation: A.
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