Mounier-Kuhn Syndrome (MKS) is a rare congenital disease that presents with abnormal enlargement in the central airways. In MKS, tracheomegaly is accompanied by difficulty in expelling recurrent lung infections and bronchiectasia. We presented a patient with MKS where commercially made stents were inadequate for stabilization and a custom-made, self-expandable metallic stent with a diameter of 28 mm and length of 100 mm was used. Chest pain that was thought to develop due to the stent and that disappeared after stent removal may be considered the main complication leading to stent removal. Continuous positive airway pressure therapy (CPAP) therapy was planned for the control of symptoms, which re-emerged after stent removal. This case is presented as an example that complications developing due to the stent as well as patient noncompliance may lead to stent removal, even when useful results are obtained from treatment of MKS.
enlargement of the airways is accompanied by collapse in expiration, resulting in symptoms. Criteria have been defined to measure dilation by tomography [8].

### Table 1: Spirometric evaluation results before and after stent insertion.

| Parameter | Before stent insertion | After stent insertion |
|-----------|------------------------|-----------------------|
| FEV<sub>1</sub> (liters) (%) | 1.43 (44.6%) | 2.95 (111.7%) |
| FVC (liters) (%) | 2.92 (72.3%) | 3.54 (114%) |
| FEV<sub>1</sub>/FVC | 0.44 | 0.83 |

FEV<sub>1</sub>: forced expiratory volume in 1 s; FVC: forced vital capacity.

tomography is shown in Figure 1. Bronchoscopy revealed an enlarged transverse diameter of the trachea starting from the entrance and severe collapse was observed with respiration. The diameter of the trachea was measured to be 35 mm at the largest dimension. Enlargement and collapse continued in the right and left main bronchus. The diameter of the airways was normal at the distal end of the main bronchus. Polysomnography was also planned with the presumptive diagnosis of obstructive sleep apnea syndrome.

Stent placement was planned for airway stabilization. Because the diameter of the trachea increased excessively, it was not preferred to use stents with the largest diameter of 22 mm because of the high risk of migration. A custom-made, self-expandable metallic stent with a diameter of 28 mm and a length of 100 mm was ordered. The stent (Silmet; Novatech, La Ciotat, France) was produced within 3 weeks. Under general anesthesia, the stent was placed in the trachea using a rigid bronchoscope (Figures 2(a) and 2(b)). Following the procedure, airway stability was improved in the trachea. The patient was transferred to the hospital ward after staying for 24 h postoperatively in the intensive care unit. There was marked improvement in symptoms. The spirometry results before stent placement and at the first-month control visit are summarized in Table 1 and bronchoscopic view is given in Figure 2(c).

At the fourth-month control visit, the patient demanded that the stent be removed due to chest pain and inability to produce sputum, which he attributed to the stent. His cardiological examination was normal and the patient stated that he did not benefit from the treatment administered for the palliation of pain. Improvement in dyspnea and exercise capacity was still maintained. In bronchoscopy, minimal secretion was observed in the stent and the stent was observed to prevent collapse markedly during expiration. Although the patient was strongly advised about the benefit of the stent, he was not persuaded and the stent was removed under general anesthesia.

After the removal of the stent, the patient was still symptomatic. At the control visit, his BMI had increased up to 43.6 and polysomnography revealed an apnea–hypopnea index (AHI) of 25.6. However, the patient could not tolerate a nasal or an oronasal mask and stated that he could not accept this alternative treatment either.

### 3. Discussion

MKS was originally defined by Mounier-Kuhn [7]. The enlargement of the airways is accompanied by collapse in expiration, resulting in symptoms. Criteria have been defined...
Figure 1: Thoracic computed tomography: in the airway segment starting from the proximal end of the trachea and extending to the main bronchi, an increase in the transverse diameter, a decrease in the anterior-posterior diameter, and diverticular irregularities in the level of the carina are observed.

Figure 2: Pictures of stent during the rigid bronchoscopy procedure. Increase in trachea transverse diameter and severe malacia (a), trachea after stent placement (b), control visit at 1st month (c).
benefit from surgical intervention [11]. Because our case refused the surgical treatment option, stent placement was attempted for symptom palliation.

Thus far, to the best of our knowledge, only one case report exists in which a custom-made large stent was used for tracheal dilation and malacia [12]. In a case with tracheomalacia associated with Marfan syndrome, due to malacia in the lower third segment of the trachea and main bronchi, a self-expandable, custom-made metallic stent with a diameter of 28 mm and a length of 69 mm was placed in the trachea and an 18-mm-diameter silicon stent was placed in the left and right main bronchi. Symptom improvement was maintained during 2 years of follow-up.

In airway stents, the formation of granulation tissue, mucostasis, infection, and migration are common complications [6, 12]; in some cases, it may be necessary to remove the stent for management of these complications. Probable complications that could develop in these patients and stent fractures could end up by the removal of the stent. For this reason, surgery should be kept in mind as a first line treatment in patients with TBM for both long term symptom palliation and not facing stent related complications. In the present case, no serious complication was observed except for chest pain attributed to the stent. Chest pain that was thought to develop due to the stent and that disappeared after stent removal may be considered the main complication leading to stent removal. Pressure exerted by the stent on a thinned tracheal wall was considered to be the main cause of retrosternal pain.

In cases with TBM, noninvasive positive pressure may help to decrease pulmonary resistance and respiratory work load and improve expiratory flow with airway stabilization and symptom control [5]. In the present case, continuous positive airway pressure therapy (CPAP), therapy was planned for the control of symptoms, which reemerged after stent removal, as well as for the treatment of obstructive sleep apnea syndrome. Because the patient did not accept CPAP either, its long-term effect on symptom palliation could not be evaluated.

4. Conclusion

In cases with severe TBM accompanied by MKS-associated tracheobronchomegaly, using large-diameter stents instead of conventional stents may be beneficial for airway stabilization and, hence, symptom control. Airway stabilization with stent can be the only option beside probable complications in cases in which traditional surgery cannot be applied. However, it should be considered that complications developing due to the stent as well as patient noncompliance may lead to stent removal, even when useful results are obtained from treatment.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

Acknowledgment

The English in this document has been checked by at least two professional editors, both native speakers of English. For a certificate, please see: http://www.textcheck.com/certificate/yCH2Mk.

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