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# An unusual phenomenon of recurrent aortic dissection with hypertension: a rare case report

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## ABSTRACT

**Introduction:** Recurrent aortic dissection is a rare and challenging phenomenon for cardiovascular cases in the world. In the last two decades among patients with initial aortic dissection, only 5% have become recurrent aortic dissection, typically seen in patients with connective tissue disorders. We report an unusual case of recurrent aortic dissection with hypertension.

**Case Presentation:** A 55-year-old female came with complaints of chest pain from the mid and left chest to the back. The symptoms had occurred for almost three years after treatment and post-surgery of her prior aortic aneurysm dissection. But she still had the same signs and symptoms as before. We requested thoracoabdominal CT angiography to confirm the diagnosis. A thoracoabdominal CTA shows recurrent aortic dissection, because of its difficulty and rarity we need proper consideration. For now, we prefer conservative medical treatment with management of blood pressure and pain control, also treatment for other diseases.

**Conclusion:** Recurrent aortic dissection is still a special challenge for physicians and a rare extreme phenomenon that may need a special approach and more complex treatment strategies.

**Keywords:** *De Bakey classification, Recurrent aortic dissection, Stanford classification.*

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## BACKGROUND

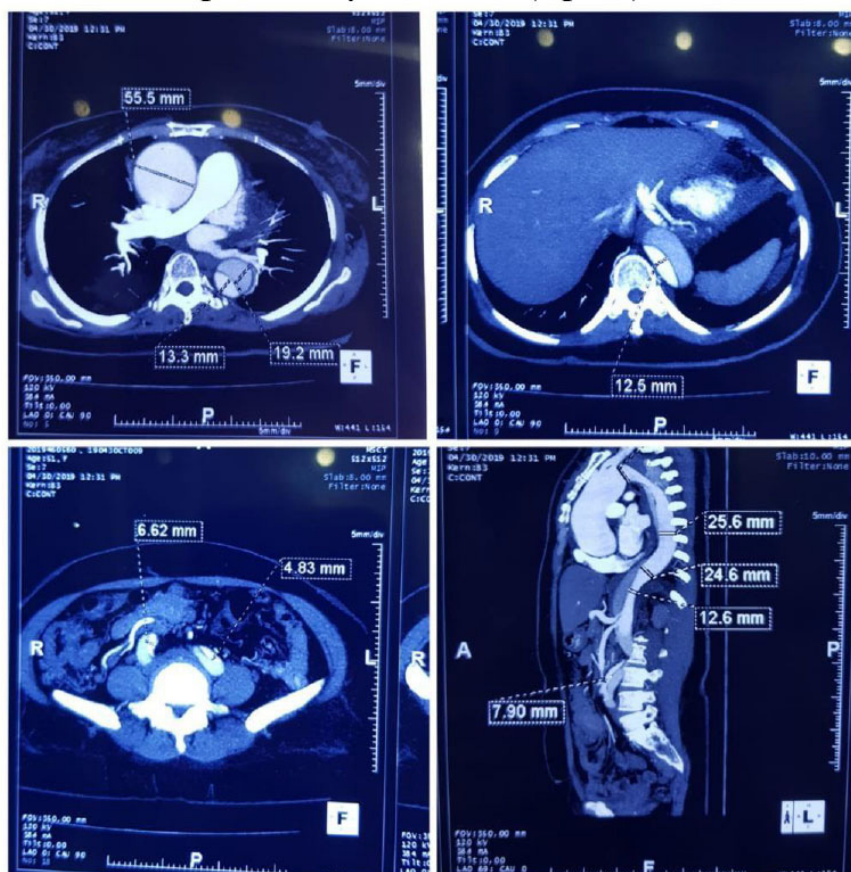
Aortic dissection is still one of the most life-threatening conditions caused by a tear in the aortic wall. In the emergency section of every 1000 patients with chest or back pain complaints, 3 of them were diagnosed with aortic dissection. The incidence of aortic dissection is 3 to 4 cases per 100.000 persons per year.<sup>1,2</sup> In specific rare incidents after treatment and surgery, new aortic dissection can reappear and become recurrent aortic dissection. Recurrent aortic dissection is a rare and challenging phenomenon for cardiovascular cases in the world. In the recent study in the International Registry of Acute Aortic Dissection (IRAD) dataset from a nearly 20-year period among 3624 patients with initial aortic dissection, only 5% become recurrent aortic dissection and typically seen in patients with connective tissue disorders, mostly in Marfan syndrome more than 21% from IRAD dataset study.<sup>3,4</sup> We report an unusual case of recurrent aortic dissection with hypertension. Consent was obtained for publication.

## CASE DESCRIPTION

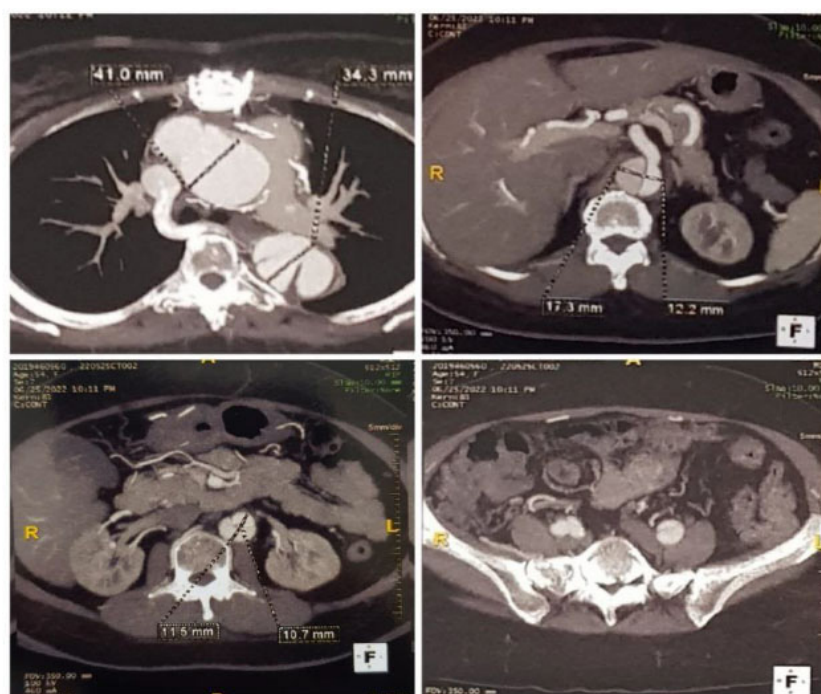
A 55-year-old female came to Bhayangkara police hospital of Jambi for a routine cardiovascular check-up with complaints of chest pain from the mid and left chest to the back like sharp and tear pain. The patient also with a history of high blood pressure. The symptoms had occurred for almost three years after treatment of her severe aortic regurgitation and ascending aortic aneurysm dissection. A thoracoabdominal computed tomography angiography (CTA) shows ascending aortic aneurysm and dissection involving descending aortic and abdominal aortic confirming the diagnosis of Stanford type B aortic dissection and type III aortic dissection according to De Bakey classification (Figure 1).

During the surgery process, it was found that an ascending aortic aneurysm and dissection starting from the aortic root, descending aortic until the abdominal aortic, confirming the diagnosis of Stanford type A aortic dissection and type I aortic dissection

according to De Bakey classification. She had a history post-surgery of her initial aortic aneurysm dissection and aortic regurgitation with Bentall procedure and hemiarch replacement with composite aortic graft in the National Cardiovascular Center Harapan Kita Hospital of Jakarta, Indonesia (December 2019). After that, the patient should undertake to check her condition every six months, but unfortunately due pandemic covid-19 she was unable to carry out therapy properly and only went to check-ups with a cardiologist in a regional hospital until 2022. However, she still had the same signs and symptoms as before, with complaints of chest pain from the mid and left chest to the back like sharp and tear pain. The patient also had any other past medical history of congestive heart failure (CHF), pericardial effusion, vertigo, hernia nucleus pulposus (HNP), and stroke. Initial examination showed blood pressure of 128/84 mmHg, heart rate of 64 beats per minute, respiratory rate was 18 breaths per minute, a temperature of 36°C, and oxygen saturation of 98% breathing ambient



**Figure 1.** Computed tomography angiography (CTA) of the aortic. Ascending aortic aneurysm and dissection flap involving descending aortic and abdominal aortic.



**Figure 2.** Computed tomography angiography (CTA) of the aortic. Dilatation of aortic arch and dissection flap involving descending aortic, abdominal aortic, until bilateral external iliac arteries.

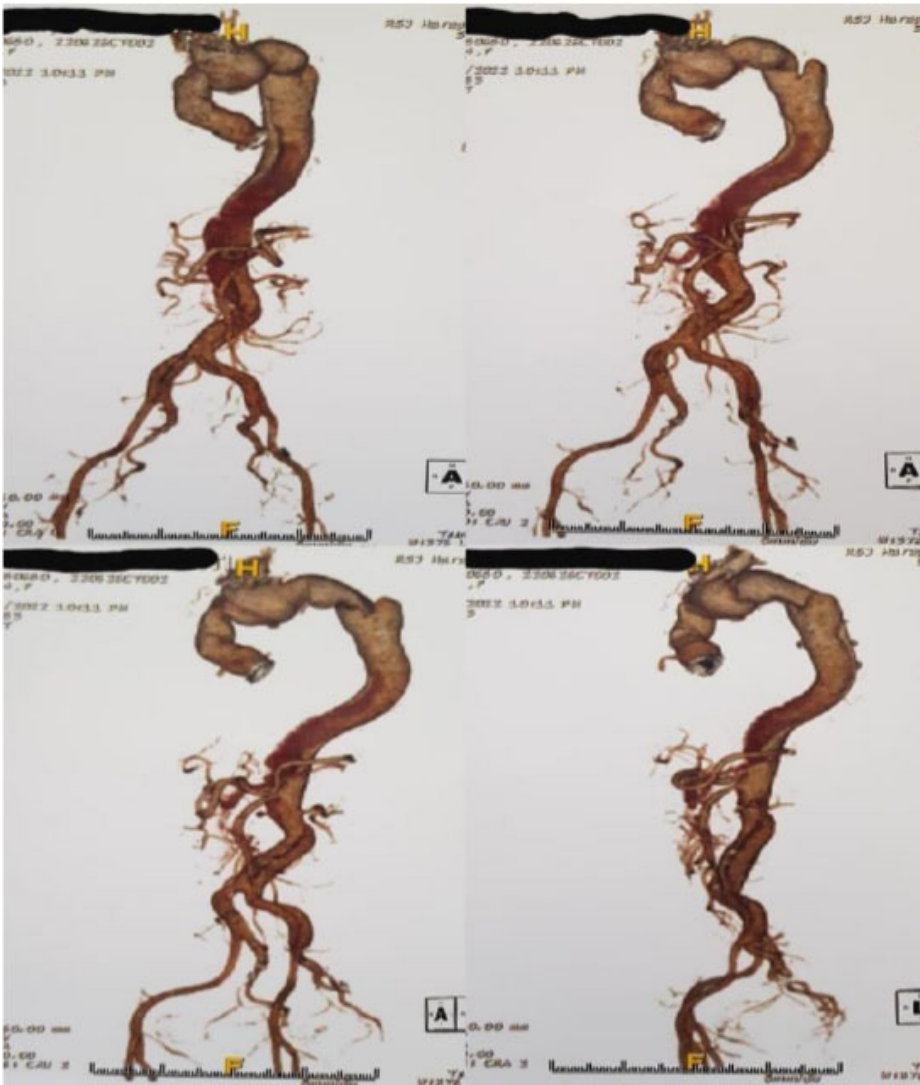
air. No abnormalities were found on examination of the heart, lungs, abdomen, and physical body.

Learn from her prior aortic dissection, the ability to mimic other condition which is difficult to diagnose properly and quickly because she had a medical history of HNP and prioritized treatment from a neurologist, on the other hand, the symptoms disguised with other cardiovascular diseases such as CHF, pericardial effusion and severe aortic regurgitation. We requested thoracoabdominal CTA again to confirm the diagnosis (June 2022). A thoracoabdominal CTA shows recurrent aortic dissection, starting from descending aortic, abdominal aortic, until bilateral external iliac arteries, confirming the diagnosis of Stanford type B aortic dissection and type III aortic dissection according to De Bakey classification (Figure 2 and 3).

Consultation and information have been given to the both patient and her family. For treatment, she gets bisoprolol, candesartan, amlodipine, hydrochlorothiazide, and warfarin. She needs to control her hypertension which is most likely to be a trigger for the aortic dissection condition dan need constant check-up with the cardiologist. Until now, no connective tissue disorders have been found in this patient and family member. Genetic testing was not performed yet because there is no indication based on her family history, but it is recommended to determine the risk factors. While waiting for the patient to return for the next check-up at the National Cardiovascular Center Hospital in Jakarta, we need proper consideration. For now, we prefer conservative medical treatment with management of blood pressure and pain control also treatment for other patients' diseases.

She was doing well and received long-term monitoring until now. Last check-up at Bhayangkara police hospital of Jambi on October 2022. The patient still had the same symptoms and sometimes her high blood pressure appeared but for now, her initial examination showed good condition. The next check-up at the National Cardiovascular Center Hospital in Jakarta is still six months later than scheduled.





**Figure 3.** The 3D reconstruction showed an intimal dissecting flap starting from the descending aortic, abdominal aortic, until bilateral external iliac arteries, confirming the diagnosis of Stanford type B aortic dissection and type III aortic dissection according to De Bakey classification

## DISCUSSION

Recurrent aortic dissection become an unusual phenomenon because of its rarity proved by a recent study of only 5 % in the world in the last two decades and only typically seen in patients with connective tissue disorders, mostly in Marfan syndrome more than 21% of IRAD dataset studies.<sup>3</sup> Difficulty occurs in this disease both in initial aortic dissection or recurrent aortic dissection. It has a wide range of clinical presentations that may mimic other conditions with chest pain symptoms, chest pain is one of the common complaints always found in the emergency room. For example, the prevalence of coronary artery disease is 100 to 200 times

more common than aortic dissection and some cases of chest pain with diagnosed acute myocardial infarction turned out to be aortic dissection.<sup>1</sup> In this case, in the initial aortic dissection the patient complains of terrific back pain with diagnosed HNP, but she also complains of constant chest pain like a tearing feeling, sometimes shortness of breath, and quickly tired. According to consideration, the patient was referred further for echocardiography and thoracoabdominal CTA, from the examination result she had severe aortic regurgitation and aortic aneurysm dissection. She got her surgery for treatment, however, she still had the same signs and symptoms as before, and

the symptoms had occurred for almost three years after treatment and became recurrent aortic dissection. Mostly all her diseases have the same signs and symptoms as her aortic dissection.

In the IRAD dataset who had documentation of aortic dissection when they were treated, from 3624 patients over a nearly 20-year period, only 204 (5%) were diagnosed with prior aortic dissection and had a recurrent aortic dissection. Initial aortic dissection patients consisted of 67% Stanford type A dissections, the recurrent aortic dissection was most likely more type B (52.5% Type B and 47.5% Type A). However, since surgery and treatment details of the first aortic dissection in the recurrent aortic dissection group are not available, we cannot assume that the type B recurrent dissections are truly new acute type B dissections and not simply residual type B dissection with the persistence of false lumen blood flow after an initial type A dissection repair, and most likely recurrent aortic dissection patients complaint no symptoms. In this case, the patient's condition is suitable according to the study, who presents with an initial ascending aortic dissection and becomes recurrent descending aortic dissection.<sup>3</sup>

Several risk factors are possible to cause aortic dissection in this case. Usually, older females affected by aortic dissection present mostly with congestive heart failure. Hypertension is considered a high-risk factor for aortic dissection in about 80% of patients with an incidence rate of 21 per 100.000 persons per year. Mostly recurrent aortic dissection is only typically seen in patients with Marfan syndrome and other inherited connective tissue disorders, such as vascular-type Ehlers-Danlos syndrome or Loeys-Dietz syndrome, especially in young patients. From a study in IRAD, more than 21% of recurrent aortic dissection had Marfan syndrome, but this patient does not have the clinical or molecular diagnosis of Marfan syndrome.<sup>5</sup> Some cases of recurrent aortic dissection have disease-causing mutations genes, for example, patients with a mutation in the ACTA2 gene. ACTA2 encodes for the actin isoform present in aortic smooth muscle, and mutations in this gene are associated with familial thoracic aortic aneurysm

and dissection disorder. Disease-causing mutations in the ACTA2 gene are the most common cause of nonsyndromic hereditary thoracic aortic aneurysm and dissection, in up to 21% of cases. Unlike connective tissue disorders like Marfan and Loeys-Dietz syndrome, patients with ACTA2 mutations usually have no physical manifestations of disease, so they may not be recognized until an aortic event occurs in the patient or a family member and genetic testing is recommended for recurrent aortic dissection case.<sup>6,7</sup> If there are no signs and symptoms of connective tissue disorders, hypertension, and older age are most likely to trigger aortic dissection, especially with a condition of aortic aneurysm.

Surgery is the definitive treatment for patients with acute aortic dissection type A, which one is works to treat or prevent complications. Successful surgery with good follow-up in 1 to 3 years provides survival rates of more than 90%. However, surgery mortality for ascending aortic dissections in the last two decades has not changed significantly, between 15% to 35%.<sup>1</sup> On the other hand, for uncomplicated acute aortic dissection type B conservative medical treatment with management blood pressure and pain control is the standard of treatment but is sometimes considered for endovascular therapy.<sup>8</sup> Thoracic endovascular aortic repair (TEVAR) is suitable for complicated acute aortic dissection type B intervention but if not suitable open surgery can be the other option.<sup>9</sup> Early diagnosis and excellent treatment are important factors for a good prognosis. Unfortunately, in this case, even though surgery has been performed on prior aortic dissection, the patient's imaging demonstrated 30 months later with a recurrent descending aortic aneurysm dissection. For a while, medical treatment with management of patients' blood pressure and pain control is still

suitable because recurrent aortic dissection in this patient is aortic dissection type B.

A wise decision is needed for this patient if the patient's condition is suitable for TEVAR, open surgery, or only needs medical treatment. Endovascular techniques continue to evolve, these patients seem possible to be treated with advanced endovascular technologies. The patient hopes her condition and disease can be treated well, she wants to know more about her particular rare case by making her case study material to help other patients with the same condition in the future with better medical treatment upgrades. So far after the treatments she received, the symptoms of chest pain and hypertension appear rarely and the frequency is reduced too.

## CONCLUSION

Recurrent aortic dissection is still a special challenge for physicians and a rare extreme phenomenon that may need a special approach and more complex treatment strategies. Due to few reports, we need more research and study on this disease to improve high diagnostic accuracy, difficult etiology, and evolving medical treatment, especially in endovascular therapy.

## CONFLICT OF INTEREST

None.

## FUNDING

None.

## ETHICAL STATEMENT

The patient has given informed consent regarding the publication of this case.

## AUTHOR CONTRIBUTION

All authors contributed equally to this study

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