Neurological Symptoms in Type A Aortic Dissections

Charly Gaul, MD; Wenke Dietrich, MD; Ivar Friedrich, MD; Joachim Sirch, MD; Frank J. Erb Guth, MD

Background and Purpose—Aortic dissection typically presents with severe chest or back pain. Neurological symptoms may occur because of occlusion of supplying vessels or general hypotension. Especially in pain-free dissections diagnosis can be difficult and delayed. The purpose of this study is to analyze the association between type A aortic dissection and neurological symptoms.

Methods—Clinical records of 102 consecutive patients with aortic dissection (63% male, median age 58 years) over 7.5 years were analyzed for medical history, preoperative clinical characteristics, treatment and outcome with main emphasis on neurological symptoms.

Results—Thirty patients showed initial neurological symptoms (29%). Only two-thirds of them reported chest pain, and most patients without initial neurological symptoms experienced pain (94%). Neurological symptoms were attributable to ischemic stroke (16%), spinal cord ischemia (1%), ischemic neuropathy (11%), and hypoxic encephalopathy (2%). Other frequent symptoms were syncopes (6%) and seizures (3%). In half of the patients, neurological symptoms were transient. Postoperatively, neurological symptoms were found in 48% of all patients encompassing ischemic stroke (14%), spinal cord ischemia (4%), ischemic neuropathy (3%), hypoxic encephalopathy (8%), nerve compression (7%), and postoperative delirium (15%). Overall mortality was 23% and did not significantly differ between patients with and without initial neurological symptoms or complications.

Conclusion—Aortic dissections might be missed in patients with neurological symptoms but without pain. Neurological findings in elderly hypertensive patients with asymmetrical pulses or cardiac murmur suggest dissection. Especially in patients considered for thrombolytic therapy in acute stroke further diagnostics is essential. Neurological symptoms are not necessarily associated with increased mortality. (Stroke. 2007;38:292-297.)

Key Words: aortic dissection More neurological complications More neurological symptoms

The incidence of aortic dissection ranges from 5 to 30 cases per million people per year.1 Aortic dissection can produce a wide range of symptoms by affecting the outflow of supracaortal, abdominal, spinal, extremity, and renal vessels. Typical symptoms are chest or back pain and hypotension leading to the differential diagnosis of myocardial infarction. Neurological symptoms occur because of occlusion of carotid, vertebral, or spinal arteries, and vasa nervorum of peripheral nerves, or because of hypotension and related cerebral perfusion deficit. Most studies that have evaluated neurological symptoms in patients with acute aortic dissection were restricted to a small number of patients at a single center. They showed a distinct increased mortality in the presence of neurological symptoms.2,3 The aim of the present study was to assess the link between type A aortic dissection and neurological symptoms with regard to perioperative and mid-term outcome.

Materials and Methods

All patients with acute aortic dissection treated in 2 departments of cardiothoracic surgery were retrospectively enrolled between January 1, 1998 and June 30, 2005. Both the Department of Cardiothoracic Surgery Halle and Nuremberg are responsible for a region of ~1 000 000 inhabitants. Patients were identified by screening hospital discharge diagnosis records and surgical databases. Clinical files of patients with thoracic or thoraco-abdominal aortic dissection fulfilling the criteria of DeBakey type I or II and Stanford type A were reviewed by neurologists (C.G. and W.D.) for neurological symptoms. The Stanford classification consists of 2 types: type A, involving the ascending aorta regardless of the entry side location; and type B, involving only the aorta distal to the origin of the left subclavian artery. The DeBakey classification consists of 3 types: type I, with involvement of both the ascending and descending aorta; type II, with involvement of only the ascending aorta; and type III, only the descending aorta.1 Neurological symptoms were classified into different groups according to Blanco et al (1999).2 Neurological symptoms of all patients admitted primarily to the emergency room or the intensive care unit of both hospitals were diagnosed after detailed clinical assessment by neurologists and further neuroimaging if necessary. In patients directly transferred from regional hospitals for further surgical management, classification was made by documented clinical signs and symptoms and neuroimaging findings. The diagnosis of aortic dissection was confirmed with CT, MRI, echocardiography, aortography, intraoperative visualization, MRI, echocardiography, aortography, intraoperative visualization,
and post-mortem examination according to the recommendations of the Task Force on Aortic Dissection of the European Society of Cardiology. Analysis included general medical history and preoperative clinical characteristics with main emphasis on neurological symptoms. Treatment and outcome were analyzed, focusing on perioperative neurological complications. Outcome of patients was determined at discharge from the acute hospital and after 6 months. The latter determination was made by contacting the rehabilitation centers or the general practitioner. Data analysis was performed using statistical analysis software SPSS 12.0. Using univariate analysis, frequency and distribution of demographic or comorbidity variables and presenting symptoms were compared in patients with and without signs of neurological dysfunction. $\chi^2$ cross-tabulations were applied as appropriate with a significance level of 0.05.

Results

Demographics, Predisposing Conditions, and Etiology

Demographics

Data of 102 patients (median age, 58 years; range, 18 to 78 years) were enrolled. Men were affected more frequently than women (1.7:1). The peak incidence in men was in the sixth and seventh decades, whereas women were affected mostly in the eighth decade.

Type of Dissection, Predisposing Conditions, and Etiology

All patients had Stanford type A dissection; DeBakey type I was identified in 60.8% of patients. The most common predisposing conditions were hypertension (in 65.3% of patients), obesity (35.6%), and cigarette smoking (31.7%). Marfan syndrome was present in 17.7% of patients (mean age, 36.7 years; range, 18 to 68 years). Histopathologic diagnostics revealed arteriosclerosis in 36.3% of the patients as the most frequent finding; in 27.5% no changes could be detected.

Initial Symptomatology

Presenting Symptoms and Signs

The majority of patients (86.1%) presented with initial chest or back pain. Major pain in the neck or head was noted in 2 patients. Among the patients with neurological symptoms at the onset of dissection, only two-thirds gave a history of pain, whereas most patients without neurological symptoms (94.4%) experienced initial pain ($P=0.001$***). Out of 102 patients, 30 (29.4%) showed neurological symptoms as the initial manifestation of aortic dissection (mean age, 57.2 years; male/female-ratio 1.3:1). DeBakey type II aortic dissection was less frequently observed among patients with neurological symptoms (23.3% versus 45.8% with/without neurological symptoms; $P=0.045$*). The observed neurological symptoms were classified as: (1) ischemic stroke (persistent or transient, e.g., stroke, or transient ischemic attack); (2) spinal cord ischemia; (3) ischemic neuropathy; and (4) hypoxic encephalopathy (Figure 1).

Ischemic neuropathy resulted from occlusion of the aortic bifurcation (N=4), iliac of femoral arteries (N=6), and subclavian artery (N=1). Typical features were signs of acute upper or lower limb ischemia as localized severe pain and paresthesia, pulse deficit, and, in severe cases, motor deficit. In contrast, acute paraplegia and complete sensory loss below level Th12 in 1 patient with no signs of lower limb ischemia was most likely attributable to spinal cord ischemia caused by aortic dissection distending from aortic valve to aortic bifurcation, occluding spinal arteries. Transient global amnesia (TGA) was identified in 2 patients. Both patients showed no pure TGA, but showed TGA-plus-syndrome with additional focal neurological signs such as mild left-side motor deficit, slight anosmia, and mild facial paresis; therefore, they were classified as having ischemic stroke. Eight of 30 patients (26.7%) presented a combination of different neurological symptoms: hemiparesis and tonic-clonic seizure or syncope (3 times), transient ischemic attack and syncope (twice), ischemic neuropathy and seizure (once), and TGA-plus-syndrome (twice). In half of all patients, neurological symptoms were only transient.

Special Neurological Diagnostics and Findings

CT scans of the brain at the time of presentation were performed in 9 patients, 8 of them with neurological symp-
toms, and as expected showed no signs of acute ischemic stroke. Doppler and duplex sonography of the extracranial arteries was performed in one-sixth of all patients (N=117), 9 of them with neurological symptoms, and revealed dissection in 5 patients.

Ischemic stroke (16 patients) was most frequently referable to the carotid circulation (N=13; 81.2%), predominantly right-sided (69.2%). Only 2 of the 9 patients with right hemispheric stroke showed unilateral dissection of the innominate artery, whereas both carotid arteries were involved in 7 patients. Involvement of one or more main branches of the aortic arch (confirmed by CT or MR angiography, carotid Duplex, or intraoperative visualization) occurred in 43.1% of all patients and 56.7% of patients with initial neurological symptoms (P=0.178; Table 1). Dissection of the innominate and carotid arteries was present in 41 patients, involvement of the subclavian artery was present in 25 patients. Preoperative stroke or transient ischemic attack was only found in 22.7% of all patients with dissection of the supra-aortic vessels.

Therapy, Complications, and Outcome

Surgical Therapy
Supracommissural replacement of the ascending aorta was performed in 46 patients (with intact or resuspendable aortic valves), whereas 52 patients (without resuspendable aortic valves) received a composite replacement. Incidence of neurological complications was similar between both groups (48.1% versus 41.3%; P=1.000).

Postoperative Neurological Symptoms and Outcome
Neurological postoperative complications were observed in a total of 47 patients (47.5%), including 17 patients with initial neurological symptoms and additional postoperative neurological complications. Complications were also divided into different groups (Figure 2). The observed nerve compression syndromes included Horner syndrome, cardiovocal syndrome or combinations of these syndromes. Eight of the 47 patients (17%) with postoperative neurological complications showed various combinations of different neurological symptoms.
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The mortality rate in the 16 patients with preoperative stroke (38.9%) was higher than that of patients without preoperative stroke (19%, \(P = 0.116\)) but did not reach statistical significance. Perioperative stroke was not related to higher mortality (26.7% versus 21.8% with/without perioperative stroke; \(P = 0.740\)). In patients with preoperative hypotension (<110 mm Hg (systolic blood pressure)) the perioperative stroke rate of 20% was higher than in patients with normotension (9.3%) but did not reach significance \((P = 0.154)\). Fifty-seven patients (72% of all survivors) were asymptomatic at discharge, 5 patients were in need of permanent care, and 14 patients were in need of help in daily life (Table 2). Although no correlation between occurrence of postoperative neurological complications and death could be observed (Table 2; \(P = 0.478\)), patients without neurological complications were more frequently independent in daily life. Significant independent determinants of perioperative or early death were (mortality with versus without presence of the predictor): preoperative hypotension (31.1% versus 13.0%; \(P = 0.047\*\)), preoperative renal impairment (58.3% versus 17.2%; \(P = 0.004\*\*\)), and history of renal insufficiency (43.8% versus 17.6%; \(P = 0.042\*\)). Logistic regression analysis showed that preoperative renal impairment and history of renal insufficiency were best predictors for hospital death: Knowledge of both variables allowed correct prediction of death in 79.6%.

Discussion

Clinical Features of Aortic Dissections

In aortic dissections, neurological symptoms are often dramatic and may dominate the clinical picture and mask the underlying condition. The frequency of neurological involvement varies from 17% to 40% (Table 3).\(^5,6\) Many neurological findings have supposedly been overlooked because of incompleteness of neurological examination in critically ill patients. Remarkably, pain is not an obligatory symptom of aortic dissection. A total of 13.9% of patients in the present study noted no pain, matching to the reported ranges of pain-free dissections between 5% and 15%.\(^6,7\) Approximately half of the patients who did not report pain solely showed neurological symptoms. Furthermore, neurological symptoms are often evanescent, fluctuating, and fully remitted before admission to the emergency room. Neurological manifestations usually appear at or soon after the onset of dissection. Rapid improvement in such cases is probably the result of only transient arterial occlusion at the moment of propagation of the dissection. Symptoms of ischemic stroke were the most common initial neurological finding. In the present series, strokes were more frequently hemispheric compared with vertebral-basilar and predominantly right sided (69.2%) matching to the frequency of 71% reported by Chase et al.\(^8\) This dominance of right hemispheric stroke despite mostly bilateral carotid dissection perhaps could be explained by different mechanical dynamics in the progression of the dissecting hematoma. Involvement of the major branches of the aortic arch was found in 43.1% of all patients in the present study, most frequently affecting the innominate and carotid arteries because of their proximity to the aortic arch.

In the preoperative stroke subgroup, the frequency of supra-aortic involvement was 62.5%, and thus clearly higher...

### TABLE 2. Outcome with Special Consideration of Initial and Postoperative Neurological Symptoms

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Asymptomatic</th>
<th>Need of Help</th>
<th>Need of Permanent Care</th>
<th>Death</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>57 (57.6%)</td>
<td>14 (14.1%)</td>
<td>5 (5.1%)</td>
<td>23 (23.2%)</td>
<td>99*</td>
</tr>
<tr>
<td>Patients with initial neurological symptoms</td>
<td>13 (44.8%)</td>
<td>4 (13.8%)</td>
<td>3 (10.3%)</td>
<td>10 (30%)</td>
<td>30</td>
</tr>
<tr>
<td>Patients with postoperative neurological complications</td>
<td>20 (45.5%)</td>
<td>10 (22.7%)</td>
<td>5 (11.4%)</td>
<td>9 (20.5%)</td>
<td>44</td>
</tr>
</tbody>
</table>

*Outcome data of 3 patients were not available.

### TABLE 3. Initial Neurological Symptoms in the Present Study in Comparison With Literature

<table>
<thead>
<tr>
<th>Neurological Symptoms</th>
<th>Present Study</th>
<th>Blanco et al(^7)</th>
<th>Álvarez et al(^2)</th>
<th>Meszaros et al(^5)</th>
<th>Fann et al(^24)</th>
<th>IRAD(^6)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of included patients</td>
<td>102 with type A dissection</td>
<td>24 with type I dissection</td>
<td>90 with type A dissection</td>
<td>75 with type A or B dissection</td>
<td>174 with type A dissection</td>
<td>289 with type A dissection</td>
</tr>
<tr>
<td>Neurological Symptoms</td>
<td>30 (29.4%)</td>
<td>6 (25%)</td>
<td>21 (23%)</td>
<td>30 (40%)</td>
<td>*</td>
<td>17%</td>
</tr>
<tr>
<td>Ischemic stroke</td>
<td>16 (15.7%)</td>
<td>2 (8.3%)</td>
<td>12 (13.3%)</td>
<td>24 (32%)</td>
<td>17 (6%)</td>
<td>(6.1%)</td>
</tr>
<tr>
<td>Spinal cord ischemia</td>
<td>1 (1%)</td>
<td>1 (4.2%)</td>
<td>8 (8.9%)</td>
<td>2 (2.7%)</td>
<td>(5%)</td>
<td>*</td>
</tr>
<tr>
<td>Ischemic neuropathy</td>
<td>11 (10.8%)</td>
<td>1 (4.2%)</td>
<td>5 (5.6%)</td>
<td>4 (5.3%)</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>Hypoxic encephalopathy</td>
<td>2 (2%)</td>
<td>2 (8.3%)</td>
<td>*</td>
<td>*</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>Syncope</td>
<td>6 (5.9%)</td>
<td>*</td>
<td>3 (3.3%)</td>
<td>*</td>
<td>*</td>
<td>(12.7%)</td>
</tr>
</tbody>
</table>

*No data available. N (%) presented.
compared with the total study population. In one-third of all patients with preoperative stroke, however, ischemic stroke was not caused by extension of the dissection toward the supra-aortic vessels, but was caused by other etiological mechanisms such as thromboembolism or severe hypotension. Furthermore, involvement of the supra-aortic vessels does not necessarily lead to cerebral ischemia. Only 22.7% of all patients with dissection of the supra-aortic vessels presented with preoperative stroke or transient ischemic attack.

Diagnosis of both acute ischemic stroke and aortic dissection is difficult when the presenting signs are atypical. Among our patients we observed a 76-year-old woman presenting with acute right hemispheric stroke and without other clear signs or symptoms of the underlying aortic dissection in whom cardiac tamponade and intrapleural hemorrhage developed after thrombolytic therapy with recombinant tissue plasminogen activator (rt-PA) and died. Only 2 other patients treated with rt-PA for acute ischemic stroke resulting from aortic dissection were previously published.9,10 Because aortic dissection is a rare cause of ischemic stroke, the aggressiveness of screening for this entity has to be questioned. An emergent chest X-ray should be considered as part of acute ischemic stroke protocols as well as paying attention to physical examination findings.11

**TGA-Like Symptoms in Patients With Aortic Dissection**

Two patients with TGA-like symptoms caused by aortic dissection were classified as ischemic stroke because of the accompanying focal symptoms. TGA-like symptoms are remarkable in 2 respects. First, TGA may extinguish important details of a patient’s recent history. One of our patients could not report the sudden onset of severe chest pain, which was the reason for the emergency call, because the memory of the episode was lost in amnesia. Second, the accompanying minor neurological deficits suggest an arterial-ischemic etiology of TGA as the result of aortic dissection.12,13 In 2 comparable, recently published patient reports, authors postulated that the acute pain of aortic dissection triggered a stress reaction responsible for TGA.14,15

Disorders of consciousness or syncopes and epileptic seizures frequently occur at the onset of dissecting aneurysm. They were present in half of all patients with neurological symptoms in the present study, consistent with literature (Table 3).6,8,16

**Operative Therapy of Aortic Dissection and Perioperative Neurological Symptoms**

Supracommissural replacement and composite replacement, each isolated or combined with replacement of the aortic arch, was applied to an equal number of patients. Matching to previous studies,17,18 incidence of neurological complications was similar between groups. The overall frequency of postoperative neurological complications (47.5% of all patients) in this series was a little higher compared with literature (Table 4), probably resulting from the exact recording and assignment by neurologists in the present study. Interestingly, more than half of patients with initial neurological symptoms showed additional new postoperative neurological complications.

The frequency of perioperative stroke (14.1%) in the present study was a little higher than previously reported but was not related to higher mortality.18,19,20 In contrast to the predominantly right-sided preoperative strokes, perioperative strokes affected equally both hemispheres and were often bilateral. Occurrence of postoperative neurological complications, especially perioperative stroke, did not correlate with supra-aortic involvement as well. This suggests that other perioperative factors such as hypothermic circulatory arrest, suboptimal cerebral protection, or microemboli seem to be responsible for perioperative stroke. Preoperative hypotension in acute type A dissection was described as a predisposing factor toward perioperative stroke (P<0.05).20 In the present study, a higher frequency of stroke in patients with preoperative hypotension was found, but this did not reach significance (P=0.154).

Postoperative confusion, agitation was observed in 14.7% of patients, comparable to the reported frequency after cardiopulmonary bypass and surgery of ascending aorta.18,19,21 The specific cause of postoperative delirium is unclear but is likely associated with subclinical microemboli or generalized cerebral malperfusion.19

There is conflicting evidence concerning the question whether presence of neurological symptoms is an independent predictor of poor outcome or in-hospital death in patients with acute type A aortic dissection. Although our study, the
IRAD,6 and Fann et al.24 revealed no such correlation, other studies identified preoperative neurological deficit as a significant predictor of hospital death.18,22,23 The overall in-hospital mortality rate in our series was 22.6%. In contrast, patients with initial neurological symptoms had a slightly, but not significantly higher mortality rate of 30%. Whereas no correlation could be observed between occurrence of postoperative neurological complications and death, patients without neurological complications were more frequently independent in daily life. These findings contrast sharply to the thesis of Álvarez Sabin, Vázquez et al, and Blanco, Díez-Tejedor et al that neurological symptoms per se would indicate an unfavorable prognosis of aortic dissection.2,3

Conclusion
Aortic dissections with neurological symptoms at onset occur in one-third of the patients without any significant pain. Additionally, in case of aphasia, unconscious or TGA patients cannot report chest pain, thus complicating the correct diagnosis. Neurologists should be alert for aortic dissection in patients presenting unusual combinations of symptoms such as involvement of central and peripheral nervous system or simultaneous occurrence of syncope, seizure, and cerebral, spinal, or peripheral nerve ischemia. Presence of neurological symptoms, even severe, does not warrant withholding surgery to the patients, because when aortic dissection is recognized early, neurological symptoms are not necessarily associated with an increased mortality.

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Disclosures
None.

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