The coral reef aorta – a single centre experience in 70 patients

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Coral reef aorta (CRA) is described as rock-hard calcifications in the visceral part of the aorta. These heavily calcified plaques grow into the lumen and can cause significant stenoses, which may lead to malperfusion of the lower limbs, visceral ischemia or hypertension due to renal ischemia. From January 1984 to February 2007, 70 patients (24 men, 46 women, mean age 59.5 years, range 14 to 81 years) underwent treatment in the Department of Vascular Surgery and Renal Transplantation, University Hospital, Heinrich-Heine-University (Düsseldorf, Germany) for CRA. The present study is based on a review of patients' records and the prospective follow-up in the outpatient clinic. The most frequent finding was renovascular arterial hypertension (44.3%) causing headache, vertigo and visual symptoms. Intermittent claudication due to peripheral arterial occlusive disease was found in 28 patients (40.0%). Seventeen

patients (24.3%) presented with chronic visceral ischemia causing diarrhea, weight loss and abdominal pain. Sixty-nine of the 70 patients (98.6%) underwent surgery; in 57 patients, aortic reconstruction was achieved with thromboendarterectomy, performed on an isolated suprarenal segment in six cases (8.7%), an infrarenal segment in 15 cases (21.7%), and the supra- and infrarenal aorta in 43 cases (62.3%). Eight patients (11.6%) died during or soon after surgery. Postoperative complications requiring corrective surgery occurred in 11 patients (15.9%). Almost one-third of the patients (n=19, 27.5%) returned for follow-up after a mean of 52.6 months (range six to 215 months). Of the 19 patients, there was significant clinical and diagnostic improvement in 16 patients (84.2%) and three patients (15.8%) were unchanged. Impairment was not observed. Despite the existing and improving surgical techniques for the treatment of CRA, its pathophysiological basis and genesis is not yet understood.

Key Words: Aorta; Atherosclerotic occlusive disease; Calcification; Coral reef aorta

Atherosclerotic occlusive disease of the aorta usually affects its distal portions, including the bifurcation, and extends into the iliac arteries. Typically, the atherosclerotic lesion begins below the renal arteries (1,2). Calcification and obstruction involving the suprarenal aorta is rare and has only been described in case reports (3-7). Coral reef aorta (CRA) is described as rock-hard calcifications in the visceral part of the aorta. These heavily calcified plaques grow into the lumen and can cause significant stenoses, which may develop into final ischemia or hypertension due to renal ischemia. The patients reported in the literature were approximately 50 years of age, which is one to two decades younger than most patients suffering from other arterial occlusive diseases. Sako et al (5) described these atherosclerotic changes of the visceral aorta as atherosclerotic occlusions of the midabdominal aorta. The frequency of this disease was estimated to range between 0.6%, or six in 1000 patients (4), and 1.8%, or two in 110 patients (5). It was not until 1984, when Qvarfordt et al (8) reported their experiences with nine patients over a period of 13 years, that this eccentric, heavily calcified polypoid lesion, originating from the posterior surface of the suprarenal aorta, was named "coral reef aorta". The macroscopic morphology – a rock-hard, irregular, gritty, whitish luminal surface – reminded the authors, who were situated on the coast of the Pacific ocean, of a coral reef (Figure 1). Therefore, the term CRA was inaugurated into the Anglo-American medical literature as a new entity of vascular disease. The case reports of CRA published in the French language called it "*Maladies obstructive calcificante de*

l'aorte thoracique descendante et de l'aorte abdominale sus-rénale" (9), meaning 'endoaortic calcific proliferation of the upper abdominal aorta' (10). Historically, the very first description of CRA can be attributed to the biographer of the Swiss anatomist Johann Jakob Wepfer (1620-1695), who mentioned the condition of Wepfer's aorta postmortem in the *Memoria Wepferiana* in 1727 (11): "...thence, everywhere, all the way to the very iliac ramifications themselves, were deposits of semi-circular shape whose consistency varied from gristle to frank bone..." (Figure 2).

Our experiences with the operative management of 21 patients treated in the Department of Vascular Surgery and Renal Transplantation, University Hospital, Heinrich-Heine-University (Düsseldorf, Germany) between 1998 and 2000 were published earlier (12). In the present paper, we describe the findings, operative procedure and follow-up of 70 patients, including clinical examination and duplex ultrasound results. We concentrated on the long-term results and the frequency of recurrent disease, with special emphasis on the visceral and renal arteries.

METHODS

From January 1984 to February 2007, 70 patients (24 men, 46 women, mean age 59.5 years, range 14 to 81 years) underwent surgical treatment for CRA. The present study is based on a review of the patients' records and the prospective follow-up in the outpatient clinic. All patients who presented with the typical signs of CRA in the noncontrast computed tomography

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Figure 1) Specimens of the disobliterated plaques in the visceral aorta – “a rock-hard, irregular, gritty, whitish luminal surface strongly resembling a coral reef” (8). **A** Intraoperative specimen; **B** Specimen before preparation for histology

examination (NCCT) and in the intra-arterial digital-subtracted angiography (DSA) were enrolled. The hyperdense depiction of calcification in NCCT allows for prompt detection of the calcification in the lumen. CRA is visualized in the NCCT as white intraluminal blocks in the visceral aorta, with a density comparable with the structure of bones (Figure 3). A cloudy formation that narrows the visceral aorta to just a very small lumen (Figure 4) is the typical morphology of CRA, as seen on the DSA. These characteristic diagnostic signs on NCCT and DSA were present in all cases. Most patients presented with the typical risk factors predisposing them to atherosclerosis. Diabetes was found rather infrequently; however, heavy smoking (up to 50 cigarettes per day) was found in 70.0% of the patients. Comorbidities such as tuberculosis, syphilis or aortitis (6,13-16) could not be detected in the study population. All patients underwent a thorough cardiological and neurological examination, including an electrocardiogram, chest x-ray, Doppler and duplex ultrasound scans of the supra-aortic branches, native CT scan of the thoracic and abdominal aorta, and DSA of the aorta and the adjacent arteries. When significant coronary artery lesions were present, angioplasty of the coronary arteries, coronary bypass surgery or surgery for carotid artery stenoses was performed before the aortic surgery. After discharge, all surviving patients and their general practitioners were contacted by telephone. The patients were encouraged to visit the outpatient clinic for a complete check-up that included filling out a questionnaire, a

Figure 2) Figure of the aorta after the postmortem of Johann Jakob Wepfer (Swiss anatomist, 1620-1695), published in his biography *Memoria Wepferiana* in 1727: “...thence, everywhere...were deposits of semi-circular shape whose consistency varied from gristle to frank bone...”. **A** Aorta; **B** Supra-aortic vessels; **C** Iliac arteries; **D** Coronary arteries; **E** Celiac trunk; **F** Renal arteries. Reprinted with permission from reference 11, copyright Elsevier Limited

thorough physical examination, a blood test and an ultrasound of the abdomen, including the kidneys and renal arteries.

Symptomatology and findings

Depending on the extent of the disease and the aortic branches involved, three leading symptoms were found, of which the most frequent finding was renovascular arterial hypertension causing headache, vertigo and visual symptoms. Thirty-one patients (44.3%) were found to take one (three patients), two (seven patients), three (eight patients) or more than three (13 patients) medications to control arterial blood pressure. All 31 patients had developed either severe renal artery stenoses or stenosis of the suprarenal aorta due to protrusion of the coral reef-like calcifications.

Intermittent claudication due to peripheral arterial occlusive disease was found in 27 patients (38.6%). The pain-free walking distance was less than 200 m (peripheral arterial occlusive disease Fontaine stage IIb) in 20 patients (74.1%) (Table 1).

Fifteen patients (21.4%) presented with chronic visceral ischemia causing diarrhea, weight loss and abdominal pain. Abdominal angina occurred in 11 of 15 patients (73.3%) and weight loss occurred in 12 of 15 patients (80.0%). Angiography of the visceral arteries showed total occlusion of branches ($n=15$; celiac trunk $n=5$, superior mesenteric artery [SMA] $n=3$, inferior mesenteric artery $n=7$) and heavily stenotic

Figure 3) Noncontrast computed tomography scan of a patient's abdomen showing massive, subtotal occluded arteriosclerosis of the suprarenal aorta

Figure 4) Digital subtracted angiography of the abdominal aorta and the iliac arteries showing cloudy gaps of contrast material in the visceral part of the aorta, typical of coral reef morphology

branches (n=34; celiac trunk n=13, SMA n=14, inferior mesenteric artery n=7) (Table 1).

Surgical procedure

Sixty-nine (98.6%) of the 70 patients underwent surgery. A 78-year-old female patient, with many comorbidities and only very mild symptoms of visceral ischemia due to an occlusion of the celiac trunk, was treated medically only. All surgical procedures were performed under general anesthesia. The operative approach was a median laparotomy in 27 patients (39.1%) and a left-sided thoracoabdominal approach in 40 patients (58.0%). In two patients, the aorta was dissected by the extraperitoneal approach. In 57 patients, aortic reconstruction

TABLE 1
Prevalent symptoms in patients with coral reef aorta (n=70)

Symptoms	n (%)	
Renal artery stenosis or arterial hypertension	28 (40.0%)	
No antihypertensive drugs	0 (0)	
One antihypertensive drug	2 (7.1)	
Two antihypertensive drugs	6 (21.4)	
Three antihypertensive drugs	8 (28.6)	
More than three antihypertensive drugs	12 (42.9)	
Peripheral arterial occlusive disease or intermittent claudication	27 (38.6)	
PAD Fontaine stage I	0 (0)	
PAD Fontaine stage IIa	2 (7.4)	
PAD Fontaine stage IIb	20 (74.1)	
PAD Fontaine stage III	4 (14.8)	
PAD Fontaine stage IV	1 (3.7)	
Chronic visceral ischemia or angina abdominalis	15 (21.4)	
Angina abdominalis	11 (73.3)	
Weight loss	12 (80.0)	
	Occlusion	Stenosis
Celiac trunk	n=5	n=13
Superior mesenteric artery	n=3	n=14
Inferior mesenteric artery	n=7	n=7

PAD Peripheral arterial disease

was achieved by thromboendarterectomy (TEA) in the layer between the tunica adventitia and the tunica media. TEA was performed in an isolated suprarenal segment in five cases (8.7%), an infrarenal segment in 14 cases (21.7%), and the supra- and infrarenal aorta in 38 cases (62.3%). In four patients (7.2%), stenosis of the aorta was insignificant and reconstruction could be restricted to the visceral and/or renal arteries only. There was one exceptional case, where the aorta was closed by a 5-0 monofilament running suture. Due to extension of the plaques into the distal aorta, a TEA of the aortic bifurcation was necessary in 33 patients (47.8%). Distal to the aortic bifurcation, a TEA of the iliac artery (unilateral n=3, 4.3%; bilateral n=24, 34.8%) and the femoral artery (unilateral n=2, 2.9%; bilateral n=2, 2.9%) was performed in 31 cases. Blood flow was restored by TEA to the celiac trunk in 29 cases (42.0%) and to the SMA in 33 cases (47.8%). Additional reconstruction was performed in eight cases (aortovisceral bypass in six patients: vein n=4, Dacron n=1, polytetrafluoroethylene n=1; transposition of SMA into the infrarenal aorta n=2) (Table 2). TEA was performed in all cases of occlusive renal artery disease regardless of the leading symptoms. Therefore, the renal arteries were reconstructed in almost two-thirds of the patients (unilateral in eight patients [11.6%], bilateral in 33 patients [47.8%]). Other simultaneous procedures (aortobifemoral bypass in five patients, thoracoabdominal graft replacement for descending thoracic aneurysm in two patients, carotid artery reconstruction before the aortic reconstruction in one patient) were carried out in eight cases. (Table 2).

RESULTS

Eight patients (11.6%) died during or soon after surgery. One patient died from acute cardiac arrest during surgery. Six

TABLE 2
Surgical procedures in patients with coral reef aorta (n=69)

Surgical procedure	n (%)
Aortic TEA	
Suprarenal aorta	5 (8.7)
Infrarenal aorta	14 (21.7)
Supra and infrarenal aorta	38 (62.3)
No aortic TEA	4 (7.2)
Direct aortic suture	50 (72.5)
Dacron patch	1 (1.4)
Iliac-femoral TEA	
Aortic bifurcation	31 (47.8)
Iliac artery	
Unilateral	3 (4.3)
Bilateral	19 (34.8)
Femoral artery	
Unilateral	2 (2.9)
Bilateral	1 (2.9)
TEA of the visceral arteries	
Celiac trunk	29 (42.0)
Including transposition on SMA	1
Including transposition on aorta-SMA bypass	1
Including vein bypass	3
Including Dacron bypass	1
SMA	33 (47.8)
Including vein bypass	1
Inferior mesenteric artery	16 (23.2)
Including PTFE bypass	1
TEA of the renal arteries	
Unilateral	8 (11.6)
Bilateral	33 (47.8)
Coprocedures other than TEA	
Aortobifemoral prosthesis	5 (7.2)
Thoracoabdominal prosthesis	1 (1.4)
Thoracic prosthesis	1 (1.4)
TEA of the internal carotid artery	1 (1.4)

PTFE Polytetrafluoroethylene; SMA Superior mesenteric artery; TEA Thromboendarterectomy

patients died of septic shock from pneumonia (postoperative day 4), myocardial and brain infarction (postoperative day 19), cardiac arrest (two patients, postoperative day 31) and multi-organ failure after a prolonged stay in the intensive care unit (three patients, postoperative days 29, 36 and 52, respectively). Postoperative complications that required corrective surgery occurred in 11 patients (Table 3). The most significant problem was acute ischemia of the lower extremities, which required inguinal TEA and/or thrombectomy in seven patients. One patient suffered from colon ischemia, which resulted in subtotal colectomy. Four patients were treated in the intensive care unit with pleural drainage due to prolonged pleural effusion. In three cases, medical treatment prevented surgical intervention (myocardial infarction, renal insufficiency and necrotizing pancreatitis). The overall rate of relevant complications was 30.4% (n=21). Late complications, leading to corrective surgery, occurred in four patients (5.8%)

TABLE 3
Perioperative mortality, morbidity and late complications with corrective procedures in patients with coral reef aorta (n=69)

Mortality (n=8, 11.6%)	Days postoperative
Acute cardiac failure	Intraoperative
Septic shock	4
Myocardial and brain infarction	19
Multiorgan failure	29
Cardiac arrest (2 patients)	31
Visceral ischemia, multiorgan failure	36
Multiorgan failure	52
Morbidity (n=21, 30.4%)	n
Acute leg ischemia	7
Pleural effusion	4
Bleeding	3
Brain infarction	2
Myocardial infarction	1
Spleen rupture	1
Terminal renal insufficiency	1
Necrotizing pancreatitis	1
Colon ischemia	1
Late complications with corrective procedures (n=4, 5.8%)	Months postoperative
Aortobiliac thromboendarterectomy	159
Aortic prosthesis, renal artery thromboendarterectomy, aortorenal PTFE bypass	89
Thrombectomy of an aortobifemoral bypass	55
Renal artery thromboendarterectomy	6

PTFE Polytetrafluoroethylene

within a time interval of six to 159 months after the first procedure. Subsequent TEA was needed in three patients: aortoiliac TEA 159 months after surgery in one patient, and TEA of the renal arteries after six months in one patient and 89 months in another patient. One patient suffered a thrombotic occlusion of the aortobifemoral prosthesis in one limb, 55 months after surgery (Table 3).

Histological findings

After prolonged decalcification of the specimen removed by TEA, a high-grade thickening of the intima became evident through hematoxylin and eosin staining; this was caused by hyalinized and partially collagenous fibres (Figure 5A). Deposits of atheromatous material containing cholesterol-crystal gaps and calcification were found as well. The inner parts of the vessel wall showed large appositions of parietal fibrin layers, with collagenous fibres originating from the intima. The thrombotic material consisted of enlarged dystrophic calcification in a coral reef-like manner (Figure 5B). Furthermore, in some specimens, the intima and media showed focal heterotrophic ossifications (Figure 5).

Follow-up

Twenty-five of the 69 patients (36.2%) died in the meantime, including the eight patients who died during the hospital stay

Figure 5) Histopathological aspects of coral reef aorta after prolonged decalcification and hematoxylin-eosin staining. **A** Evidence of hyalinized and partially collagenous fibres. **B** Thrombotic material consisting of enlarged coral reef-like dystrophic calcification

postsurgery. Two patients died from a malignant disease (metastatic esophageal cancer and pancreatic cancer, respectively), two patients died of renal failure and three patients died of vascular complications (myocardial and brain infarction, and visceral ischemia, respectively). In 10 cases, the cause of death could not be identified due to follow-up times of up to more than 20 years. Twelve patients of the surviving 44 surgical patients (27.3%) were excluded from follow-up because of a prohibitively short postoperative period. Thirteen patients were lost to follow-up or refused to come to the hospital for re-examination because of advanced age and/or the long distance to travel to Düsseldorf. Almost one-third of the patients (n=19, 31.1%) came to the outpatient clinic appointment for follow-up. The follow-up interval ranged from six to 215 months postsurgery, with a mean of 52.6 months. The follow-up examination included medical history, a physical examination and a Doppler or duplex ultrasound scan of the kidneys, including the resistance index.

Compared with the patients' conditions before surgery, there was significant clinical and diagnostic improvement in 16 of the 19 follow-up patients (84.2%). Three patients remained stable (15.8%). In all cases, there were no observed clinical or diagnostic signs of impairment. All 10 patients suffering from arterial hypertension were still on antihypertensive medication, but with reduced medication – two patients took one medication, four patients took two medications, two

TABLE 4
Follow-up of 19 patients concerning their primary symptoms

Symptoms	n (%)
Renal artery stenosis or arterial hypertension	10 (52.6)
No antihypertensive drugs	0 (0)
One antihypertensive drug	2 (20)
Two antihypertensive drugs	4 (40)
Three antihypertensive drugs	2 (20)
More than three antihypertensive drugs	2 (20)
Peripheral arterial occlusive disease or intermittent claudication	5 (26.3)
PAD Fontaine stage I	1 (20)
PAD Fontaine stage IIa	4 (80)
PAD Fontaine stage IIb	0 (0)
PAD Fontaine stage III	0 (0)
PAD Fontaine stage IV	0 (0)
Chronic visceral ischemia or angina abdominalis	4 (21.0)
Angina abdominalis	0 (0)
Weight loss	0 (0)

Follow-up duration of 6 to 215 months postoperative, mean 52.6 months. PAD Peripheral arterial disease

Figure 6) The number (No) of antihypertensive drugs used before and after surgery, and during follow-up, shows a significant decrease. One-way ANOVA using SPSS (SPSS Inc, USA), $P < 0.05$; Bonferroni's multiple comparison test, $P < 0.0001$

patients took three medications and two patients took more than three medications. The level of creatinine was normal in eight patients and slightly elevated in two patients. Five patients underwent surgery for intermittent claudication. Four of them could walk more than 200 m, and one patient was not at all limited in walking. The four patients with history of chronic visceral ischemia were in good physical condition and did not experience problems with digestion or weight loss (Table 4). Statistical evaluation was performed by SPSS (SPSS Inc, USA) (one-way ANOVA, $P < 0.05$; Bonferroni's multiple comparison test, $P < 0.0001$). The only significant parameter was the number of antihypertensive drugs (Figure 6).

Ultrasound of the kidneys

Kidney size was measured with an ultrasound scan. The mean size was 10.0 cm (range 7.0 cm to 11.8 cm) for the right kidney and 10.4 cm (range 7.5 cm to 12.1 cm) for the left kidney. These values were consistent with the normal range. The calculated resistance index, which was also within the normal range, was 0.69 (range 0.48 to 0.80) for the right kidney and 0.68 (range 0.48 to 0.81) for the left kidney. Because the measurement of the resistance index is a relatively recent diagnostic standard in sonography, and has only been available for the past 10 years, a full pre- and postoperative follow-up could not be completed in those patients who underwent surgery more than 10 years ago.

DISCUSSION

Since its inauguration in 1984 by Qvarfordt et al (8) (Table 5), CRA has become a widely used term. The articles published before 1984 described the same vascular disorder as acquired constructive lesions of the distal thoracic aorta (17), intraluminal calcification of the abdominal aorta above the renal arteries (8) or calcified aortic plug syndrome (18). The sum of all single case reports and small series of patients with CRA found in a MEDLINE-based search produced 41 patients (6,8-10,17-29). Qvarfordt et al (8) reported nine women between 48 and 67 years of age (median 51 years); men were not included in this series. The published experiences of the remaining 32 patients showed a median age of 48.6 years (range 27 to 79 years) and were therefore in a comparable range with the data presented by Qvarfordt et al. However, unlike the observations of sex distribution reported by Qvarfordt et al, these studies had a nearly equal sex distribution: 55.3% women and 44.7% men. The mean age in the present study was 59.5 years (range 14 to 81 years) in a population of 46 women (66%) and 24 men (34.3%). The sex distribution in the present study, with 66% women, lies within the reported range of 54.5% to 100%.

The literature review of 41 CRA patients revealed several significant clinical observations: intermittent claudication was a leading complaint in 50.0% of patients, renovascular hypertension in 41.7% of patients and chronic visceral ischemia in 9.1% of patients. One patient underwent surgery for blue toe syndrome due to recurrent embolic calcified plaques from the CRA (23). These numbers are comparable with the present findings.

Open TEA of the occluded parts of the aorta and transaortic TEA of the aortic branches are two of the most widely used surgical procedures for CRA. Flynn and Kattus (19) were the first to publish their results on aortic TEA in 1959. He removed a calcified aortic plug extending from the descending aorta into the renal arteries in a 27-year-old woman suffering from renovascular hypertension. As a result, her blood pressure values decreased and remained within the normal range. In 34 (82.9%) of the 41 CRA patients described in the literature, laparoscopic TEA was performed twice (29), two patients were treated by interposition after partial resection of the aorta (one homograft [17] and one Dacron prosthesis [9]) and four patients were treated by an aortobiiliac bypass without TEA. Concomitant procedures, other than aortic TEA, were necessary in most of the cases. Sixteen patients received bypasses (aorto-aortic = 5, aortobiiliac = 2, aortofemoral = 5, aortorenal = 2, aortomesenteric = 2), five patients received TEA of a visceral branch (renal artery = 5, SMA = 2, celiac trunk = 1). In

the present patient group, there was a higher incidence of adjunctive surgical procedures.

The recent reported experiences of the postoperative course and follow-up intervals are very inhomogeneous. Three of the 41 CRA patients died perioperatively (intraoperative death [7], multiorgan failure on day 2 and pulmonary dysfunction on day 14 postoperatively [8]). The perioperative mortality amounted to 7.3%, in comparison with 11.6% in the present series of 69 surgical patients. The follow-up period for 30 patients ranged from several weeks to nine years, and was described as satisfactory. Diagnostic evaluation during follow-up was not stated in any of the reported cases.

Apart from CRA, there is another vascular disorder of the same segment of the aorta called middle aortic syndrome (15). Sen et al (15) reported a group of 16 patients (12 women and four male patients between the ages of three and 30 years), whose chief complaint was hypertension. The pathological correlate turned out to be aortic stenosis due to inflammatory hypertrophy of the adventitia and intima layer of the vessel wall. Only one patient presented with diffuse calcification of this aortic segment. Remarkably, all patients previously had tuberculosis; thus, tuberculosis aortitis was presumed to be the cause of tissue changes in the aortic wall (15). Six of the 16 patients underwent sympathectomy, TEA or bypass surgery. Two case reports were found on patients with middle aortic syndrome at a more advanced age (61 years [30], 54 years [31]). Bialkowski et al (16) reported angioplasty and stenting of the thoracic aorta in a 13-year-old girl with Takayasu's arteritis. Severe heart failure and arterial hypertension were identified as the clinical counterpart of the aortic stenosis. The two-year follow-up showed a patent stent but a progression of the arteritis despite continuous immunosuppression.

The pathophysiological basis of CRA is still not understood. Attempts to understand this vascular entity are based on singular observations of comorbidities in patients suffering from CRA, such as syphilis (13), neurofibromatosis (32), rubella (6,33) and local, not generalized, amyloidosis (24). The patients in our group did not suffer from any of these diseases. A different approach to understanding the pathology of CRA is the assumption that exogenous trauma, such as rapid deceleration in a car accident, may lead to slight dissections inside the aortic wall. Over time, an extensive reparation process in the dissected vessel wall may trigger this excessive calcification (6,34). In the present patient group, there was a 17-year-old girl who had a bicycle accident in her childhood. Some authors assume that Takayasu-arteritis, because it is an inflammation of the vessel wall, may lead to CRA (6,14,15). However, this assumption has not yet been confirmed. The hypothesis of the influence of parathyroid hormone and serum calcium levels as possible initiators of the calcification process in the aorta has been ruled out (8,12).

A new concept has become the centre of interest in our patient collective. Patients suffering terminal renal insufficiency are, after long-term hemodialysis, at a higher risk for cardiovascular disease, such as myocardial or brain infarction, than patients without renal disorders. The common risk factors for cardiovascular diseases, such as smoking and arterial hypertension, are not sufficient to explain the pathogenesis of the sclerosis in CRA patients (35,36,37). Attempts to comprehend metabolism and regulation of calcium led to discovery of a serum protein, which was named α_2 -Heremans-Schmid glycoprotein, or fetuin-A. There is evidence that lack of fetuin-A in

TABLE 5
Literature regarding patients with coral reef aorta (CRA) and its synonyms

Author	Year	Term	Patients	Symptom(s)	Therapy	Outcome
Cooley and De Bakey (17)	1954	Acquired constrictive lesions of the distal thoracic aorta	33-year-old M	Hypertension	Aortic resection, homograft 9 cm long	Repeat procedure with graft lengthening 4 months later, healthy 4 months later
Flynn and Kattus (19)	1959	Coarctation of the aorta with calcific atheromatous degeneration	27-year-old W	Hypertension	Aortic TEA (first description)	Healthy 1 year later
Sen et al (15)	1963	Middle aortic syndrome	16 patients (12 W, 4 M, 3–30 years of age)		5 patients underwent surgery	
Lipchik et al (7)	1964	Obstruction of the abdominal aorta above the level of the renal arteries	45-year-old M	Hypertension	TEA expected	Intraoperative death due to cardiac arrest
Sako (5)	1966	Arteriosclerotic occlusion of the midabdominal aorta	53-year-old M	PAD	Aortic TEA	Healthy 1 year later
Charnsangavej (6)	1981	Intraluminal calcification and occlusion of the abdominal aorta above the renal arteries	46-year-old M 38-year-old M	PAD Hypertension	Aortic TEA Aortic and renal TEA, aortoaortic bypass	Healthy Normotension postoperatively
Walter et al (18)	1981	Calcified aortic plug syndrome	38-year-old W 69-year-old W 79-year-old W 54-year-old W 49-year-old W	Hypertension Hypertension, PAD PAD Hypertension Hypertension, PAD	Conservative Conservative TEA aorta, RA, Ce tr, SMA Aortoaortic, birenal and mesenteric bypass Aortoiliac bypass, renal revascularization	– – Healthy Healthy Healthy
Qvarfordt et al (8)	1984	Coral reef atherosclerosis	9 W (48–67 years of age)	5 patients PAD 2 patients visc ischemia 2 patients hypertension	9 patients aortic TEA 5 patients aortofemoral bypass	2 patients died (postoperative day 2, MOF; postoperative day 14, pulmonary insufficiency) 7 patients fit and well (1 to 9 years)
Bergqvist et al (20)	1985	Obstructive suprarenal aortic lesions	62-year-old W 47-year-old W	Hypertension Visc ischemia	Aortic TEA Aortic TEA	Healthy (6 years) Healthy
Patra et al (9)	1985	<i>Maladie obstructive calcifiante de l'aorte thoracique descendante et de l'aorte abdominale sus-rénale</i>	45-year-old M 57-year-old M	Hypertension Hypertension, PAD	Aortic resection, Dacron interposition Aortoaortic bypass	Healthy (4 years) Healthy (15 months)
Borgis et al (21)	1986	<i>Korallenriffarteriosklerose</i>	66-year-old W	Hypertension	No operative procedure because of poor general condition	
Fichelle et al (38)	1986	<i>Arthériosclérose oblitérante de l'aorte abdominale haute</i>	– –	Hypertension, PAD Hypertension, PAD	TEA TEA	Healthy Healthy
Peillon et al (10)	1989	Endoaortic calcific proliferation of the upper abdominal aorta	40-year-old M 55-year-old M 56-year-old M	Hypertension, visc ischemia Hypertension PAD	TEA of aorta, SMA, RA TEA of aorta, SMA, RA Aortobiliac bypass	Healthy (2 years) Healthy (6 months) –
Oberstein et al (22)	1990	<i>Korallenriff-Arteriosklerose</i>	56-year-old M	PAD	Aortobiliac bypass	–
Rosenberg and Killewich (23)	1995	Coral reef aorta	45-year-old W	Blue toe syndrome	Aortic TEA	Healthy (3 months)
Combe et al (24)	1997	Coral reef infrarenal aorta	37-year-old W	PAD	Aortoaortic bypass	Healthy (18 months)
Cappeller et al (25)	1998	<i>Korallenriffatherosklerose</i>	55-year-old W	Hypertension, PAD	TEA RA, aortoaortic bypass	Healthy (5 weeks)
Minnee et al (26)	2005	Coral reef aorta	3 patients	–	–	–
Dinis da Gama et al (27)	2006	Coral reef aorta	53-year-old W	–	AIB, bypass to visc arteries	Healthy (2 months)
Teebken et al (28)	2006	Coral reef aorta	58-year-old W	PAD	AIB	Healthy (30 months)
Di Centa et al (29)	2006	Coral reef aorta	46-year-old M 48-year-old M 52-year-old W	PAD PAD PAD, visc ischemia	Total laparoscopic removal of CRA and AIB (n=2) or open thoracic TEA (n=1)	Healthy (38 months) Healthy (29 months) Healthy (1 month)

AIB Aortobiliac prosthesis; Ce tr Celiac trunk; M Man; MOF Multiple organ failure; PAD Peripheral arterial disease; RA Renal artery; SMA Superior mesenteric artery; TEA Thrombendarterectomy; W Woman; Visc Visceral

vivo may contribute to the development of extraosseal calcification. Therefore, fetuin-A has some importance in the inhibition of ectopic calcification. Patients on chronic dialysis have a fetuin-A deficiency, and this may be a possible explanation for the increased incidence of cardiovascular disorders in these patients (35). The potential correlation between the lack of fetuin-A and the pathogenesis of CRA is still under investigation.

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