Myocardial Thievery: The Coronary-Subclavian Steal Syndrome

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Coronary-subclavian steal syndrome entails the reversal of blood flow in a previously constructed internal mammary artery coronary conduit, which produces myocardial ischemia. The most frequent cause of the syndrome is atherosclerotic disease in the ipsilateral, proximal subclavian artery. Although coronary-subclavian steal was initially reported to be rare, the increasing documentation of this phenomenon and its potentially catastrophic consequences in recent series suggests that the incidence of the problem has been underreported and that its clinical impact has been underestimated. We review the causes and background of coronary-subclavian steal; methods of preventing, diagnosing, and treating it; and the potential influence of various treatment regimens on long-term survival and the likelihood of late adverse events in patients with coronary-subclavian steal syndrome.

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Coronary-subclavian steal (CSS) was once believed to be rare, but the increasing documentation of this phenomenon and its potentially catastrophic consequences suggests that the incidence of CSS has been underreported and that its clinical impact has been underestimated. In this study, we review the causes and background of CSS; methods of preventing, diagnosing, and treating it; and the potential influence of various treatment regimens on long-term survival and the likelihood of late adverse events in patients with CSS syndrome.

Background

Coronary-subclavian steal syndrome is defined as a reversal of flow in a previously constructed internal mammary artery (IMA) coronary conduit, producing myocardial ischemia. Typically, CSS syndrome is caused by proximal subclavian artery stenosis in patients with an ipsilateral IMA coronary conduit. The anatomic findings, physiologic alterations, and pathologic consequences were initially described by Harjola and Valle [1] in 1974. In their report on a patient who presented with angina after an otherwise successful coronary artery bypass grafting (CABG) procedure, the authors stated that the anatomic, physiologic, and clinical associations among their findings represented a distinct clinical entity. Within 3 years, others applied the name “coronary-subclavian steal syndrome” to the process after recognizing the similarities between the pathologic mechanisms of this syndrome and those of the previously recognized vertebral-subclavian steal syndrome that produces vertebrobasilar insufficiency [2, 3].

As initially described more than 30 years ago, proximal subclavian artery stenosis, which is the most common cause of CSS, is usually caused by atherosclerotic disease [1–3]. However, several other pathologic processes can also compromise the subclavian artery flow to cause CSS, including Takayasu’s arteritis [4, 5], radiation arteritis [6], and giant cell arteritis [7]. Furthermore, absence of a proximal subclavian artery stenosis does not preclude the occurrence of CSS; the reversal of IMA coronary conduit flow and the subsequent myocardial ischemia that comprise the CSS syndrome have been reported to occur in association with an upper-extremity hemodialysis fistula [8–10] and an anomalous connection of the...
left subclavian artery to the pulmonary artery in d-transposition of the great arteries [11].

Presentation
Although CSS has significant consequences, its clinical impact was not fully appreciated until recently. Initial reports suggested that most patients with primary and recurrent CSS experience angina [12–16]. However, recent reports of CSS cases increasingly note “silent” ischemia [17], congestive heart failure [16, 18–20], ischemic cardiomyopathy [20, 21], and myocardial infarction [22, 23] as presenting cardiac symptoms. Therefore, it is likely that early cases of the most severe manifestations of CSS syndrome went unrecognized, leading to an underappreciation of both the incidence and the potential clinical impact of CSS [20]. In patients with known coronary artery disease and a history of coronary revascularization, sudden death or myocardial infarction is likely to be attributed to the more commonly occurring progressive, native-vessel coronary artery disease rather than to unrecognized or progressive great vessel disease that can produce ischemia. Sullivan and colleagues [23] recently reported that of their 27 patients with CSS, 16 (59.3%) presented with stable angina and 11 (40.7%) presented with unstable coronary syndromes, including 4 acute myocardial infarctions.

Incidence
Although an early study by Brown [2] suggested that the incidence of significant brachiocephalic disease in patients who undergo elective CABG is 0.5% to 2.0%, a more recent study reports that the incidence of concomitant disease is 0.1% to 0.2% [20]. Nonetheless, failure to identify significant subclavian disease before placement of an ipsilateral IMA coronary conduit or to recognize the progression of brachiocephalic disease after placement of an ipsilateral IMA coronary conduit may lead to the development of CSS and myocardial ischemia [24]. Initial reports detailing the causes and clinical manifestations of CSS described the problem as rare [3, 24]. However, several recent reports involving larger numbers of patients show that CSS is more common than was initially appreciated (Table 1). These findings suggest that in a busy cardiothoracic program, approximately 2 to 4 cases per year may be found [16, 19, 20, 23, 25–33].

Several factors contributed to an early underestimation of the incidence of CSS. These factors included the less frequent general use of IMA conduits during the time period that followed the recognition of CSS as a distinct clinical entity, the initial lack of development and subsequent low availability of noninvasive methods for diagnosis of concomitant brachiocephalic disease, the failure to apply effective diagnostic methods, a limited awareness of the problem, the lack of effective preoperative screening for concomitant brachiocephalic disease before CABG, and a tendency to attribute the most severe manifestations of CSS syndrome to other causes. The documentation of larger numbers of patients with CSS during the last 10 years (Table 1) reflects an increased awareness of the problem and the application of more effective diagnostic methods.

Screening and Diagnosis
The identification of anatomic findings that may lead to CSS, the physiologic alterations of early CSS, and the pathologic changes of CSS syndrome will significantly influence the management and outcome of patients in several clinical settings. These include patients about to...
undergo elective CABG, patients who develop clinical symptoms after CABG, and patients who have been treated previously for known CSS.

In patients about to have elective coronary revascularization, significant subclavian or innominate artery disease is most easily recognized if an upper extremity blood pressure differential is detected [34]. However, such a differential is not always found in these patients, because 31% of patients with brachiocephalic disease also have significant multivessel disease that may affect pressures in both upper extremities [35]. Alternative screening methods that may produce more useful information include ultrasonic duplex scanning before and after arm exercise, and direct angiography, which can be performed during evaluation of the coronary arteries [36, 37] and to perform arch aortography and four-vessel cerebral angiography if significant subclavian artery disease is found. This practice has enabled us to identify several patients with great vessel disease before they underwent elective CABG [20, 38]. It has also enabled us to document in several patients the progression of brachiocephalic atherosclerosis from a nonsignificant disease to a radiologically and clinically significant one that causes CSS after an otherwise successful coronary revascularization.

Angiography also produces the most efficient screening and diagnostic information for symptomatic patients who have had previous CABG or a prior intervention for known CSS. The use of angiography in such patients adds minimal risk when performed concomitantly with cineangiography, which is needed to rule out progressive native vessel coronary artery disease or inadequate conduit flow in these patients.

In the evaluation of asymptomatic patients after intervention for known CSS, noninvasive imaging using the method of Grosveld and colleagues [37] provides reliable screening information. Ultrasonic duplex scanning of the brachiocephalic and vertebral vessels, as well as hemodynamic measurements and neurologic examinations, are performed before and after exercise to differentiate hemodynamically significant lesions from nonsignificant ones. Extremity exercise is standardized (1.5 W/sec for 5 min) by means of an ergometric apparatus. An examination is considered positive if any of the following are detected: (1) new or recurrent vertebrabasilar, hemispheric, or coronary (related to CSS syndrome) symptoms or signs; (2) a decrease of 0.15 or more in the blood pressure index of the ipsilateral arm compared with the contralateral arm; (3) reversal of flow in the ipsilateral vertebral or carotid artery; or (4) common carotid artery evaluation indicating a marked serial increase in velocity (> 130 cm/s), a marked serial decrease in the ratio of blood flow velocity in the internal carotid artery to that in the common carotid artery. Although neck exposure is limited, direct visualization of occlusive or ulcerated proximal plaque is occasionally possible. Before ultrasound technology was incorporated, screening and follow-up examinations of these patients relied solely on hemodynamic and clinical evaluation. A positive examination leads to aortic arch and four-vessel cerebral angiography in each case.

Noninvasive imaging techniques, including high resolution, multi-slice computed tomographic scanning and magnetic resonance angiography, have shown promise in some recent studies when used to screen for and diagnose brachiocephalic disease [5, 10]. Although the applicability of these techniques is limited by their cost and availability, each method is noninvasive and may provide a dynamic picture if used in combination with duplex assessment of intraluminal velocity [39]. In addition, contrast-enhanced, color-coded, real-time sonography is currently being assessed and has been shown to clinically differentiate blood flow direction [40].
Brachiocephalic Manifestations

Although patients with CSS syndrome may have a broad spectrum of symptoms related to myocardial ischemia, the same great vessel disease that causes CSS may also produce concomitant symptoms unrelated to the heart (Fig 1). Proximal subclavian artery disease may also affect the posterior cerebral circulation, the ipsilateral brachial circulation, or both. Therefore, compromise of subclavian artery flow may cause the simultaneous occurrence of CSS and vertebral-subclavian steal, producing posterior cerebral insufficiency and vertebrobasilar symptoms [41–45]. This same distribution of disease may also compromise distal flow, resulting in brachial insufficiency and extremity claudication, or produce emboli, resulting in signs of extremity microembolization [46].

The finding of concomitant, multivessel brachiocephalic disease is common [35]. Significant atherosclerotic disease in the common carotid artery, innominate artery, or contralateral subclavian artery may produce concomitant symptoms related to the respective circulations supplied by those vessels. Common carotid artery disease may compromise blood flow or produce emboli that affect the anterior cerebral circulation, resulting in neurologic symptoms. Innominate artery disease, if present, may affect the right subclavian artery circulation, the right common carotid artery circulation, or both [14, 38].

If any of the previously mentioned patterns of neurologic change occur after coronary revascularization (whether or not the patient has cardiac symptoms), the clinician should consider the possibility that CSS is present. Detection of CSS at this early stage would enable intervention before the potential onset of ischemic symptoms. Although symptoms may appear early if an IMA conduit was constructed in the presence of unrecognized, ipsilateral brachiocephalic disease, they more commonly appear years after surgery as the brachiocephalic disease progresses from mild to severe. Bryan and colleagues [19] report that among 5 patients with post-CABG CSS syndrome, mean time from surgery to the report of symptoms was 7.8 years (range, 1 month to 18 years). Similarly, Elian and associates [27] report that symptoms associated with CSS began an average of 5.8 years (range, 1.7 to 10.5 years) after CABG in the 7 patients in their series.

Management

Although CSS primarily affects the heart and produces heart-related symptoms, CSS can be treated effectively with noncardiac interventions. Successful correction of CSS with relief of symptoms has been accomplished by aortic-subclavian bypass [20, 38]; carotid-subclavian bypass [19, 47, 48]; axillo-axillary bypass [49]; transposition of the IMA [50]; and percutaneous transluminal angioplasty with stenting [16, 25, 26, 28, 51, 52]; laser ablation [53], rotational thrombectomy [54], or atherectomy [55] of the subclavian artery. Although subclavian stenosis has been found to recur after several types of intervention [16, 20, 25, 56–58], the excellent long-term patency after bypass remains the standard to which the outcomes of all other procedures are compared. Multiple studies have shown that a 10-year actuarial patency of more than 90% with acceptably low morbidity and mortality can be achieved by using either direct reconstruction methods that preserve aortic inflow when multiple great vessels are involved [21, 41, 59, 60] or extra-anatomic, cervical bypass for single-vessel brachiocephalic disease [61–63]. Excellent results have been achieved using Dacron (Hemashield; Meadox Medicals, Inc, Oakland, NJ) or polytetrafluorethylene conduits [21, 59–63].

As a less invasive alternative to operative bypass, endovascular interventions for the treatment of recurrent and primary CSS are being evaluated at several institutions [16, 23, 25, 26, 62]. Early results have shown acceptable operative and early patency, although midterm patency rates are somewhat lower than those associated with operative bypass [23, 26, 62]. Endovascular intervention offers tangible benefits regarding cost, level of invasiveness, and subjective patient satisfaction [62]. For these techniques, however, we do not know yet the long-term durability, patterns of failure, efficacy as an adjunct to CABG, anticoagulation requirements, efficacy as treatment for complex (multivessel) disease, or long-term cost. Until these additional questions are answered, the precise indications for endovascular intervention versus operative reconstruction as treatment for brachiocephalic disease remain unsettled. Long-term results will provide further insight into the advantages and disadvantages of the endovascular approach and provide further direction regarding optimal management of primary and recurrent CSS.

Little is known about the natural history of untreated or medically managed CSS. However, Bryan and colleagues [19] report observational and medical management of one patient who refused percutaneous or invasive intervention for CSS. This patient expired less than 12 months after the initial diagnosis.

Clearly, if preoperative testing shows significant subclavian artery disease ipsilateral to a planned IMA coronary artery conduit, CSS may be avoided by using all-vein coronary conduits. This approach to treating patients with coronary artery disease and concomitant brachiocephalic disease is used at several institutions [64] and is associated with acceptably low operative morbidity, low operative mortality, and excellent long-term brachiocephalic graft patency in patients receiving concomitant reconstruction. Nonetheless, the use of all-vein conduits in such patients has become controversial [16, 20, 25, 65], because a recent study showed that patients who undergo concomitant brachiocephalic reconstruction and CABG using all-vein conduits have a poor 10-year actuarial freedom from death and adverse cardiac events [20]. Although choosing not to use the IMA as a coronary conduit effectively eliminates the possibility of CSS, it also denies the patient the proven benefits of IMA conduit use, which include increased long-term survival and a reduced incidence of adverse cardiac events [66]. The demonstrated safety of concomitant brachiocephalic reconstruction and CABG and the excellent long-term patency of the brachiocephalic vessels after reconstruc-
tion have led several groups to initiate studies of concomitant ipsilateral subclavian artery reconstruction and CABG with use of IMA coronary conduits in patients whose concomitant disease is recognized preoperatively. The investigators hope to improve long-term survival and decrease the incidence of late, adverse cardiac events after surgery [16, 20, 25, 65].

Although the lack of an IMA conduit in patients having concomitant brachiocephalic reconstruction and CABG may increase the likelihood of late, adverse cardiac events, it is likely that these late events are influenced by several factors, including the systemic distribution of atherosclerosis. Therefore, lifestyle change and risk-factor modification are aggressively encouraged in these patients. Long-term rates of adverse events and survival continue to be closely followed in these patients to determine whether this management approach improves outcome.

As an alternative to the use of either all-vein conduits or an IMA conduit after brachiocephalic reconstruction in patients with concomitant disease, the use of free IMA grafts or radial artery conduits may be considered. However, the specific long-term outcomes associated with these conduits and their relative impact on freedom from late, adverse cardiac events compared with that of IMA grafts have not been definitively established. Decisions about the operative approach, technique, and staging to be used with an individual patient must ultimately be based on the institution’s experience, the patient’s particular risk factors, and the severity of the disease.

We currently do not use endovascular treatment for brachiocephalic disease in conjunction with CABG, because the open surgical treatment of brachiocephalic disease has proven safe and durable at our institution, and the need for CABG negates the advantages of angioplasty and stenting that relate to cost, level of invasiveness, and need for anesthesia. However, we do use the endovascular technique in patients who have CSS after CABG. These patients are followed as part of an ongoing study that maintains strict follow-up with serial duplex examinations before and after surgery [37].

Follow-Up

Close follow-up is essential for all patients who receive brachiocephalic reconstruction as treatment for CSS syndrome. The wide spectrum of symptoms and modes of presentation that may accompany recurrent disease, the unproven long-term utility of specific intervention types, and the possibility that a catastrophic outcome may be the initial mode of presentation warrant serial clinical and noninvasive examinations [34, 67]. Performing serial duplex examinations before and after arm exercise has been reported to effectively diagnose recurrent brachiocephalic disease [37, 68]. Although this diagnostic method requires a trained technician, it has the advantages of high sensitivity and specificity, noninvasiveness, and low cost [37, 68, 69]. Other direct and indirect methods that have been used to establish the diagnosis of recurrent CSS include arch aortography, cerebral angiography [25], and sestamibi imaging [70].

Comment

In summary, CSS syndrome is a distinct clinical entity that may have profound consequences for the survival and lifestyle of the individual patient. Increased awareness of the problem and screening for brachiocephalic disease in patients with coronary artery disease will help to decrease the incidence of CSS. Preoperative angiographic screening limited to brachiocephalic vessels proximal to a potential IMA conduit is an effective screening method for concomitant disease and adds little risk when performed concomitantly with cineangiography of the coronary vessels. However, the most appropriate way to manage concomitant brachiocephalic disease and coronary artery disease is unsettled. Studies are currently ongoing in which the IMA is used as a coronary conduit after brachiocephalic reconstruction of the ipsilateral subclavian artery in patients with concomitant disease. Patients who have CSS syndrome after CABG have been successfully treated with both operative bypass and endovascular techniques. Close follow-up is essential after treatment of CSS syndrome in order to decrease the likelihood of adverse and catastrophic events that may be associated with recurrent disease.


